

CERTAIN CONGENITAL ANOMALIES:
SOME PSYCHO-SOCIAL IMPLICATIONS IN ADULTHOOD

JOAN INGRID HEYDENRYCH

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ABSTRACT

This study is an investigation of some psycho-social implications in adulthood of being born with a congenital anomaly. The congenital anomalies - oesophageal atresia, Hirschsprung's disease and high anorectal malformations - are surgically corrected at birth, but can be associated with residual problems. These problems could put patients at risk for psycho-social maladjustment.

The three anomaly groups were seen to represent varying degrees of severity. The oesophageal atresia respondents represented the no to mild disability/residual problems group. Those who had Hirschsprung's disease represented the moderate disability/residual problem group. The high anorectal malformation respondents' represented the severe disability/residual problem group. The research hypothesis is that the severity of residual problems and psycho-social functioning will be directly proportional to each other, ie. the more severe the handicap, the poorer the psycho-social functioning.

A research study was conducted on 38 adult patients whose congenital anomalies were surgically corrected at The Red Cross War Memorial Children's Hospital. The research methods used were a descriptive survey method and a case-study method. The former involved three self-administered questionnaires. Information obtained concerned demographic, socio-economic, family background, medical and psycho-social problem data. An in-depth case-study was conducted with one respondent from each anomaly group. Information was obtained concerning the effect that residual problems had had on various aspects of patients' lives. Data was analysed descriptively.

The findings of the study supported the research hypothesis, the medical prognosis and on the whole agreed with the literature. Severity of residual problems was found to be directly related to psycho-social functioning. Patients with severe disability/residual problems were experiencing the most psycho-social problems, those who had moderate disability/residual problems were found to have some psycho-social disability/residual problems, whereas those with mild disability/residual problems were found to have few or no psycho-social problems.

Self-esteem, depression, interpersonal relationships and restricted social functioning were the psycho-social aspects found to be most affected by residual problems.

The study revealed gaps in both medical and social work services for these patients in terms of ongoing follow-up services. Recommendation to improve these services have been proposed.

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CHAPTER 1

INTRODUCTION

1.1 BACKGROUND

According to Smithells (1978), surveys show that 2-2,5% of all babies born after the 28th week of gestation have birth defects sufficiently severe to cause serious handicap or to require surgical correction.

As medical science in general improves, so too has the specialized field of paediatric surgery. Following surgery many children born with anomalies are now able to live, and are furthermore being restored to functional normality or near normality. However, there are those patients who suffer from residual problems which could threaten their psycho-social functioning.

Recently, De Wet (1984) conducted a study at The Red Cross War Memorial Children's Hospital (to be referred to as The Children's Hospital) examining the psycho-social effect on the family of having a child with a congenital anomaly. This study revealed that the majority of patients with Hirschsprung's disease and anorectal malformations had been subjected to peer ridicule and ostracism at school, as a result of their socially embarrassing symptoms.

Although in some cases residual problems may have been eliminated, it seems likely that where there are problems, there may be a range of difficulties affecting the patient's psycho-social functioning.

In view of the above findings, a study focusing on the psycho-social effects of the congenital anomaly on the patient himself/herself was seen

as a worthwhile pursuit. The Department of Paediatric Surgery, Red Cross War Memorial Children's Hospital (Cape Town, South Africa), has always had a keen interest in congenital anomalies and a great deal of medical research has been conducted with the anomalies under discussion. (Louw and Cywes 1967; Louw et al 1971; Louw, Cywes et al 1971; Cywes et al 1971; Davies et al 1975; Cywes 1975; Cywes et al 1976; Davies et al 1979). Although not the most common congenital anomalies seen at The Children's Hospital, a number of cases pass through the hospital each year.

However, the importance of studying the particular anomalies under discussion is not associated with their frequency. Rather it is because they were each seen to represent a different degree of severity of residual problems related to the congenital anomaly. For this reason and as a follow-up to De Wet's (1984) study, the present study was conducted to examine the following congenital anomalies - Hirschsprung's disease, high anorectal malformations and oesophageal atresia. These three anomalies represent different degrees of severity of residual problems after surgical correction. In general, oesophageal atresia patients are expected to have no or only a few residual problems (mild severity). Hirschsprung's disease patients are expected to have some residual problems initially, but they tend to diminish as the patients get older (moderate severity). High anorectal malformation patients may have major residual problems (extreme severity). In general it is hypothesized that the severity of residual problems and psycho-social functioning will be directly proportional to each other, ie the more severe the handicap, the poorer the psycho-social functioning.

This study therefore proposes to examine the psycho-social effect in young

adulthood of people born with various congenital anomalies, which required corrective surgery during childhood leaving the patient with no or mild, moderate or severe residual problems or disability.

1.2 RATIONALE FOR THE STUDY

The writer became interested in the topic under discussion while working in the Department of Paediatric Surgery as a research social worker. As discussed, her predecessor had studied the effects on the family of having a child with a congenital anomaly. After consultation with the Head of the Department, Professor Cywes, the writer decided that a useful follow-up study to the above would be to examine the effects on the patient of being born with an anomaly. Results of the study could have implications for paediatric surgery in terms of management and follow-up procedures.

Social workers have been involved with the physically handicapped and chronically ill for a long time and have been concerned with the psycho-social effects of illness/disability. Results of the study could have implications regarding counselling of patients with regard to coping with psycho-social difficulties at different ages.

It was decided that the study should focus on adults. Adulthood is the time when one can best assess the psycho-social effects of being born with an anomaly. As Mattsson states '... the final outcome of the child's attempts at mastering the continuous stress associated with his disability cannot be assessed until young adulthood' (Mattsson 1972:805).

1.3 AIM OF THE STUDY

The aim of this study is to examine the long-term psycho-social implications, if any, of being born with an anomaly with reference to the

patient. As mentioned, the anomalies chosen for the present study were oesophageal atresia, Hirschsprung's disease and high anorectal malformations. It aims to explore the broad issues involved in having residual symptoms from a congenital anomaly, both in the past and at present (with emphasis on the latter). This would indicate the most important areas for future research that could focus on more specific aspects.

A. Specific aims

- (a) to determine if there is a relationship between severity of disability and psycho-social functioning. If so, to identify the nature of this relationship.
- (b) to explore broad areas in a patient's life most affected by residual problems.
- (c) to identify similarities and differences between anomaly groups in regard to needs and problems.
- (d) to identify any present physical problems relating to the original anomaly and/or the associated surgery and provide assessment and/or treatment and/or referral.
- (e) to make recommendations to medical and social work staff regarding follow-up of patients in the area of physical and psycho-social functioning, based on the needs and problems identified.
- (f) to make recommendations for future research and specify the areas in which this research is required.

1.4 DEFINITION OF TERMS

The following definitions concern the most important concepts in this study. Definitions of other medical terms are contained in a glossary based on the Dorlands Medical Dictionary (24th Ed.1974).(see Appendix 1)

A. Congenital anomaly

"Congenital" is defined as 'present at birth' (Warkany 1971:3).

"Anomaly" is defined as 'marked deviation from the normal standard, especially as a result of congenital or hereditary defects' (Dorlands Medical Dictionary 25th Ed. 1974:100). Therefore congenital anomalies are defects present at birth.

"Malformation" is defined as 'defective or abnormal formation; deformity' (Dorlands Medical Dictionary 25th Ed. 1974:904) and

"abnormality" is defined as 'the quality or fact of being abnormal. A malformation or deformity' (Dorlands Medical Dictionary 25th Ed. 1974:3). The terms anomaly, abnormality and malformation are seen as synonymous in this study and are used interchangeably.

B. Psycho-social

"Psycho-social" is defined as 'that which is constituted partly of psychic and partly of social phenomena' (Dictionary of Sociology 1964:240). Hollis in the Encyclopedia of Social Work defines psycho-social approaches as pertaining to 'both the inner psychological realities of man and the social context in which he lives' (National Association of Social Workers 1971:1217).

C. Adulthood

"Adulthood" is defined as 'to grow up' (Chambers Twentieth Century Dictionary 1972:17). It refers to the age when a person is seen to have matured. There is a controversy over the age at which adolescence ends and adulthood begins. The World Health Organization defines adolescents as those people aged between 10 and 19 years (McKay 1983). For the purposes of this study, adulthood refers to persons of 20 years old and over.

D. Oesophageal atresia

"Oesophageal atresia" is defined as 'congenital lack of continuity of the esophagus, commonly associated with tracheoesophageal fistula ...' (Dorlands Medical Dictionary 25th Ed. 1974:162).

E. Hirschsprung's disease/aganglionosis

"Hirschsprung's disease" is defined as 'massive enlargement of the colon, resulting from obstruction caused by an aganglionic segment of bowel' (Dorlands Medical Dictionary 24th Ed. 1965:431). Aganglionic refers to the absence of ganglion (nerve) cells.

F. Anorectal malformation

"Anorectal" is defined as 'pertaining to the anus and rectum or to the junction region between the two' (Dorlands Medical Dictionary 25th Ed. 1974:101). Therefore anorectal malformations refer to abnormalities of the anus and rectum (see p 29 for an explanation of high anomalies).

1.5 EXPLANATION OF TERMS AND ABBREVIATIONS

For the purposes of this study the meanings of the following terms and abbreviations apply:

OA Oesophageal atresia

HD Hirschsprung's disease

AM Anorectal malformations (high anomalies)

'While growing up' and 'Past' refers to the respondent's life up to the age of 19 years

'Present' refers to the respondent's life from the age of 20 onwards

'Respondents' refers to patients who took part in the present study.

1.6 CONCEPTUAL FRAMEWORK

An exploratory study aims at investigating a field that has not been previously examined. Therefore it is not clear initially what concepts and issues are going to be important when studying a certain research issue. The following is a brief description of some functional concepts that were

seen to be potentially useful in examining the topic under discussion at the outset of the study.

A. Hospitalization, separation and surgery

As the patients in the study were all subjected to surgery, hospitalization and separation from parents, it is seen as useful to look at these concepts.

Hospitalization, with the accompanying separation and loss can be viewed as a crisis situation (Sheridan 1975). It can be very stressful as it involves an unfamiliar environment, separation from parents and various, often painful, medical procedures (Robertson 1958). Traumatic separations in childhood from one or both parents can have adverse effects (Robertson 1958) and could affect the child's ability to form relationships later in life (Bowlby 1960).

Many studies note the traumatic effect of surgery which reactivates the universal fears of having one's body mutilated and views of medical procedures as punishment for actual or imagined wrongs (Mattsson 1972). Theories of bodily integration and castration anxiety are well documented in the literature (Blotcky and Grossman 1978; Berg and Berg 1983). However Korsch (1975) suggests that it is difficult to assess the specific emotional impact of a surgical procedure in very small infants, as it is not known how pain is perceived by these infants. However, she notes that separation from parents is a very distressing experience. Rutter (1979) points out that forced separations of mother and infant in the neonatal period may damage parent/child relationships. Bonding between parents and children may be further strained if there is a great deal of technological equipment. In later childhood, hospitalization can

disrupt the child's life (Sheridan 1975) eg. peer relationships, schooling.

B. Developmental stages

The psycho-social effects of illness and the associated surgery, hospitalization, separation from parents, friends, and so on, needs to be considered within the context of the developmental age of the patient and the particular life tasks with which he/she is dealing. Reference will be made to two of the most important developmental theories: Freud's psycho-sexual stages and Erikson's psycho-social stages (as described in Hall and Lindzey 1978). These theorists describe development in terms of various stages in which certain issues are important and certain tasks have to be mastered for healthy psycho-social development.

The stages are summarized as follows:

(a) First year of life

In this stage described by Freud as the 'Oral' stage and Erikson as the 'Basic Trust vs Mistrust' stage, consistent regular satisfaction of needs and bonding with the mother or caretaker is important for the child to develop trust, optimism and warmth. Separation from parents (in the case of hospitalization) and feeding problems (in oesophageal atresia patients) may have negative long-term consequences (eg. affect the parent/child relationship or other relationships later in life).

(b) Second year

This stage is described by Freud as the 'Anal' stage and Erikson as the 'Autonomy vs Doubt' stage. Here assertiveness and physical control is important for the child to develop a sense of autonomy, pride and accomplishment otherwise he/she may be

dependent on parents and have an inability to be assertive. At a stage where physical control of one's body is important an illness that undermines that control, eg. Hirschsprung's disease, anorectal malformations, may have a disruptive effect on psycho-social development. Hall and Lindzey (1978) point out that a sense of loss of self-control can cause lasting feelings of shame and doubt.

(c) Third to fifth year

Described by Freud as 'Phallic' and Erikson as 'Initiative vs Guilt' the child's exploratory behaviour and self-initiated activities at this stage are important to develop conscience, self-worth and goal definition. Children still struggling with basic physical tasks, eg. toilet training, may have difficulty at this stage.

(d) Sixth year to puberty

Freud's 'Latency' stage and Erikson 'Industry vs Inferiority' stage involve co-operation and competition from the child to develop competence, mastery of skills and self-confidence. Children who are still incontinent, who are often absent from school and therefore miss school work and who cannot take part in sport, may struggle at this stage. This may make them feel very inadequate which could have negative consequences (eg. lower their self-esteem).

(e) Adolescence

At this stage described by Freud as 'Genital' and Erikson as 'Identity vs Role Confusion', one's identity is integrated and one develops a sense of continuity with one's past, present and future identity. An adolescent who is still suffering from socially embarrassing symptoms at an age where being attractive to

the opposite sex and being part of a group are important, may find this stage difficult.

(f) Early adulthood

Erikson describes this stage as 'Intimacy vs Isolation' (Freud's stages only go up to adolescence) and here it is important to care deeply for another person and develop vulnerability. This helps one to form stable commitments and close relationships and people still suffering from embarrassing symptoms or from sexual or fertility difficulties (which can occur in anorectal malformations and Hirschsprung's disease), may find this age most stressful.

The adults in the present study were studied at the early adulthood stage of their life and this will therefore be examined further.

Gerdes et al (1981) view early adulthood as the stage when a person is emotionally independent of his parents, when he is able to support himself and when he accepts the responsibilities of an occupation, marriage and parenthood. They cite certain tasks which are associated with this age and these include selecting a mate, learning to live with a marriage partner, becoming a parent and rearing children, establishing and managing a home, becoming established in an occupation and developing a pattern of recreation. Most of the patients can accomplish these tasks, but some of them with residual problems may have difficulties in that their occupational choice and recreational activities may be restricted. They may experience interpersonal relationship difficulties and/or marital difficulties. Within the marriage sexual and fertility problems may arise.

C. Defaecation - taboos and societal reaction

Two of the congenital anomalies under discussion are associated with defaecation difficulties, i.e. anorectal malformations and to a lesser degree Hirschsprung's disease, and this concept will be briefly examined.

Defaecation is a normal bodily function which is probably talked about less than any other bodily function. There seems to be a universal taboo surrounding elimination of waste matter. Defaecation, as a process, and faeces, as a product, are generally discussed in a humorous context as the subject of jokes or comical situations. It is usually seen as an impolite or offensive topic and generally avoided.

Douglas (1966) in examining concepts of purity and pollution notes that some cultures see dirt avoidance as a matter of hygiene and aesthetics and some see it as an aspect of a religion. Furthermore, there is a great variation in the way in which different aspects of bodily functions are treated in different parts of the world. Each culture has various rituals and beliefs associated with defaecation, eg. in the Islamic culture one hand is used for eating and one for cleaning oneself and if a person steals his eating hand is cut off, so that he has to eat and wipe himself with the same hand, which is seen as very humiliating.

Kelly (1986) points out that bowel disorders carry a particular symbolic significance in Western cultures which could lead to a tendency for patients and their families to engage in collusion to minimize the symptoms or deny the severity of the condition.

One must take into account the potential consequences of having bowel problems in a society where this is seen as an offensive, secret and taboo matter.

D. Concepts of stigma, handicap and sick role

Some of the patients with Hirschsprung's disease and many patients with anorectal malformations may have been stigmatized due to the socially embarrassing nature of their illness and might also have been in a sick role for part of their lives.

All these concepts are associated with one's self-concept and body image. Self-concept may be described as a person's view of his own attributes and Gerdes et al (1981) see this as including physical, psychological and social attributes. The evaluative aspect of self-concept is self-esteem, which is to a certain extent determined by others' evaluation of oneself. A negative evaluation, especially in childhood and adolescence where self-esteem is often determined by others, may have serious implications for psycho-social functioning. Sensky (1982) notes that stigma is described by Goffman as shameful differentness. Sensky (1982) notes that the handicapped are not considered responsible for their handicaps, yet when it comes to coping with the stigma, the onus is on the stigmatized rather than others. He also notes that successful management of stigma is often judged in terms of the reduction of such discomfort. Referring to the patients under discussion, it is their socially embarrassing symptoms, eg. odour associated with defaecation difficulties that must be managed and covered up, rather than others accepting and 'putting up' with the symptoms.

Another important concept is that of a sick role and the associated behaviour which Mechanic (1962) describes as 'illness behaviour'. Sensky (1982) notes that those with a congenital physical handicap are often assigned a 'sick role' or 'role of helplessness'. Bracht (1978) comments on Parson's concept of sick role and notes that when a person's illness is legitimized by medical sanction, that person occupies a special role in society. People who have been sick from birth may be affected by the evaluation of others, and by themselves in their sick role.

E. Coping

Reif (1973) points out that the main concern of a person who is chronically ill is basically the same as that of the healthy individual - managing his personal, social and occupational activities as well as possible. She notes that a person with a chronic disease must function despite a disease which may have a profound effect on his/her life. She makes an important point that medical intervention is not sufficient for the person to lead a normal life. Various coping strategies have to be employed.

Barofsky (1981) reduces various coping strategies to three broad categories (1) techniques to minimise distress (2) activities that attempt to deal with specific issues and (3) activities that involve others, eg. self-help groups.

The first strategy is seen as an important one for the patients in the present study. One finds patients involved in behaviour aimed at avoiding certain situations or feelings, controlling events and detaching themselves from potentially upsetting situations. Reif (1973) points out that this means patients have to modify or curtail

some activities, for example social activities, drastically. Barofsky (1981) notes that there is a controversy over the relationship and distinction between 'defences' and 'coping'. Defences are generally seen as negative and maladaptive, while coping is seen as positive and adaptive. Some authors see defences as part of the coping process (Barofsky 1981).

Kelly (1986) gives a personal account of his experience with a bowel disease - ulcerative colitis. The effects of these symptoms are similar in some ways to Hirschsprung's disease and anorectal malformations. His main methods of adjustment were the processes of denial, normalization and accommodation to the disease, which are factors common to many illnesses. He notes that many authors have commented on the danger of strategies like denial and normalization, while others have seen them as healthy adaptive strategies to very real problems. Kelly takes the latter view and states '... normalization and denial may involve actions which are in and of themselves socially or psychologically deviant. However, the deviance, ironically, helps to preserve, on the surface and in public encounters, a sense of normality' (Kelly 1986:661). He feels that this is important in that if these strategies or defences are broken down, the patients may be forced into openly deviant roles, eg. sick persons or invalids.

Wiener (1975), in her discussion of rheumatoid arthritis patients, compares the normalization process with that of ulcerative colitis patients. She notes that the normalization strategy of covering up is easier for rheumatoid arthritis patients as pain is invisible, whereas the external evidence of diarrhoea in ulcerative colitis

patients is visible. Covering up strategies for the latter patients are very difficult.

Reif (1973) identifies some key features of chronic illness - the symptoms interfere with many normal activities and routines, the medical strategies are limited and there is a substantial disruption of daily living. She notes that most of the time a person with ulcerative colitis is concerned with the social and psychological consequences of the symptoms. As the symptoms are often very socially embarrassing, as are those of Hirschsprung's disease and anorectal malformations, various practical strategies are often employed to conceal the problem.

The tactics include (1) preventing symptoms, eg. diarrhoea, from occurring in the first place (preventive strategies) eg. regulating diet, (2) keeping odour and excrement from being noticed by others (protective strategies) eg. planning outings according to accessibility of toilets and (3) correcting the damage or disruption if an 'accident' occurs (corrective strategies) eg. change of clothing.

It is obvious that coping involves various practical and psycho-social strategies which will differ from patient to patient.

The concepts described above will be examined further in the discussion (of findings) section in the context of the research hypothesis.

1.7 OVERVIEW OF THE STUDY

The following chapter (Chapter 2) provides an overview of congenital anomalies in general and the specific anomalies under discussion. The next chapter (Chapter 3) reviews the literature pertaining to the psycho-social effects of illness/disability. Medical and social work resources and facilities available to patients (born with the congenital anomalies under discussion) are briefly examined (Chapter 4). A description follows of the research methods used in the study (Chapter 5). In the next chapter, the main findings are presented (Chapter 6), followed by a discussion of these findings in the context of the research hypothesis and the literature (Chapter 7). Lastly, conclusions concerning the study are drawn and various recommendations are made based on these conclusions.

CHAPTER 2

CONGENITAL ANOMALIES (Anomalies in general and the specific anomalies under discussion)

2.1 ANOMALIES IN GENERAL

As already discussed, (in Chapter 1 p 5) the terms anomaly, abnormality and malformation will be used interchangeably.

A. Background History - beliefs and aetiology.

For centuries men and women from all over the world have been fascinated by monstrous or deformed people. The film 'The Elephant Man' based on a true story about a man with gross physical deformities was a recent box office success. Throughout time, abnormal people have been popular subjects or characters in books and plays; dwarfs are still commonly seen in comic roles in circuses.

Warkany (1971) notes that, in the past, when children with birth defects were born it aroused people's emotions and they reacted with admiration, awe or fear. Deformed people were either killed or worshipped. Sculptures, carvings and drawings of abnormal births show the early knowledge and interest in rare and unusual humans.

Regarding aetiology, a range of superstitions and theories existed. These included beliefs that malformations were forms of punishment for sins that had been committed, that they were the result of associations between witches and humans, that they were caused by mating between animals and humans (the hybridity theory) or that they were the result of parents' mental impressions at the time of

conception or during pregnancy, eg. mothers frightened by rabbits produced children with hare-lips.

As scientific knowledge has increased many of the ancient superstitions have disappeared. However, it is important to bear these beliefs in mind as many birth defects are not satisfactorily explained. If parents of congenitally malformed children are presented with inadequate explanations, they tend to develop their own theories and explanations, some of which may be based on past superstitions. Furthermore, one must bear in mind the effect that parental ideas and beliefs will have on the child. At some stage the congenitally malformed child will probably ask his/her parents questions concerning the aetiology of his/her anomaly.

B. Present beliefs and aetiology

Very little is known about the causation of congenital abnormalities. Smithells (1978) sees them as being the result of a sequence of events. Concepts of causation include genetic factors, chromosomal changes, molecular changes, maternal diseases (viral, metabolic or nutritional) or drugs taken in pregnancy, but there is still a lack of consensus regarding aetiology. A few of the major aetiological theories will be described briefly.

(a) Genetic and chromosomal factors

Regarding genetic factors, some congenital malformations are transmitted as hereditary characteristics and can be attributed to genes (dominant or recessive) inherited from one or both parents. There are various dominantly inherited congenital malformations, eg. extra finger anomalies and autosomal

recessively inherited congenital malformations, eg. hydrocephaly. Warkany (1971) notes that over 150 malformations are transmitted in this way. Some congenital malformations are caused by sex-linked inheritance and over 30 malformations are seen to be transmitted in this manner, eg. imperforate anus.

Regarding chromosomal factors, many congenital malformations, especially multiple defects, are seen in persons who have chromosomal abnormalities. The best known change of this kind involves duplication of an entire chromosome.

(b) Environmental factors

Warkany (1971) notes that interference with normal pre-natal development between conception and birth is usually seen as environmental in contrast to those of genetic origin, although the latter and chromosomal abnormalities are also attributed to environmental causes. Environmental influences, eg. infections, chemical or mechanical insults leading to vascular interference at different stages of pregnancy can influence the child's formation in different ways.

Maternal infections eg. rubella, are well-known in the causation of various congenital abnormalities, eg. ocular, cardiovascular, aural and mental defects. Various drugs taken by the mother during pregnancy can cause congenital abnormalities, the most well-known incident being the introduction of thalidomide in 1956 as an influenza remedy and a sedative which produced an epidemic of children born with severe limb deformities. Another environmental cause is foetal irradiation which has been linked

with many congenital malformations, eg. spina bifida, clubfoot, cleft palate, etc. Finally some maternal disorders have led to congenital abnormalities. For example iodine deficiency was associated with endemic goiter and cretinism and was a major health problem in certain areas where the soil, water and vegetation lacked iodine. Other maternal disorders, eg. diabetes have been associated with congenital malformations or increased perinatal mortality of the child (Warkany 1971).

It is obvious from the above discussion that the aetiology of congenital malformations has not been clearly established and may be due to multiple causes or causes not yet identified. In fact, Smithells (1978) notes that multifactorial defects probably account for a quarter to one-third of all congenital defects.

C. Classification of congenital anomalies

Warkany (1971) classifies congenital malformations into malformations of:

- the central nervous system
- the eye
- the ear
- the endocrine glands
- the cardiovascular and respiratory system
- the face
- the oro-gastrointestinal tract
- the systemic skeletal system
- the axial and appendicular skeleton
- the muscular system
- the urogenital system

- the skin, as well as
- tumour-associated malformations

The abnormalities under discussion are all part of the oro-gastrointestinal tract malformations and will be discussed further.

2.2 SPECIFIC ANOMALIES (oesophageal atresia, anorectal malformations, Hirschsprung's disease).

It is important when examining the psycho-social impact of the congenital anomalies under discussion to know exactly what the congenital anomaly/illness is, what the symptoms are and what residual problems may be expected.

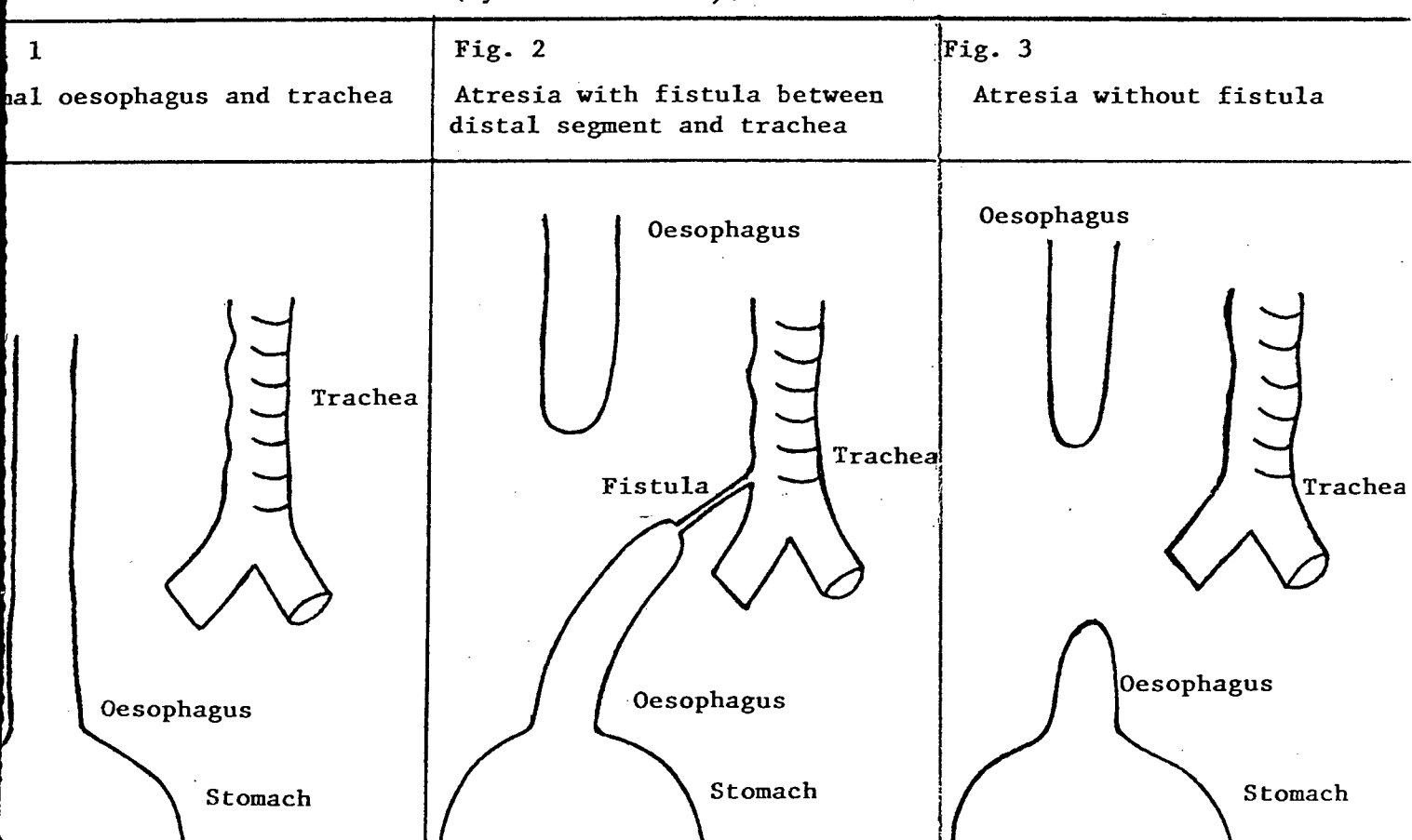
All the anomalies under discussion are life-threatening and require major surgery. When the respondents in the present study were operated on, the hospital had recently opened and surgery for these anomalies was in its infancy. Respondents often had to spend a great deal of time in hospital, and had repeated operations especially where post-operative complications occurred. The knowledge pertaining to the pathophysiology of these anomalies has increased and surgical techniques has improved considerably. At present patients usually have fewer post-operative complications and therefore spend less time in hospital than did the respondents under discussion. It must be noted that some patients spend more time in hospital than they need to because poor socio-economic conditions are not conducive to the very ill post-operative patient. One must bear in mind the potential effect that long periods of hospitalization, separation from parents, repeated operations, painful, uncomfortable and in some cases, embarrassing residual symptoms may have on a person's psycho-social functioning.

A. Oesophageal stresia

This anomaly was first described by Durston in 1670 (Randolph 1986) with the usual form ie. associated with a fistula of the trachea, described by Gibson in 1696 (Cywes et al 1976). Successful operations were only performed in the 1900's with the first survivor being operated on in 1939 (Cywes et al 1976).

(a) Pathology

Oesophageal atresia refers to a congenital gap in the oesophagus. This results in the oesophagus ending in two blind pouches. As noted, this anomaly is usually associated with a fistula with the trachea and Cywes et al (1976) estimate that this occurs in over 90% of patients who have this anomaly. There are various anatomical variations of this anomaly, but the most common are (1) an atresia with a fistula between the distal segment and the trachea (see Fig.2). This occurs in 85-90% of patients and the second most common is (2) an atresia without a fistula with a long gap between the two blind ends of the oesophagus (see Fig.3). This occurs in 7-9% of cases (Cywes et al 1976).



(b) Aetiology

The aetiology of this anomaly is unknown and although familial cases have been reported no true genetic influences have been found (Cywes et al 1976; Cudmore 1978). Other studies indicate that oesophageal atresia is more likely to be associated with a developmental disruption due to embryolic and environmental factors (Randolph 1986).

(c) Associated anomalies

More than half the children presenting with tracheo-oesophageal fistulas have associated anomalies, especially cardiac and rectal anomalies (Cudmore 1978; Warkany 1971; Cywes et al 1976). Approximately half of these are major anomalies which require urgent surgical correction. Other anomalies include renal tract, cardiac, vertebral and skeletal anomalies. The survival rate of children with oesophageal atresia is significantly affected by the presence of other anomalies (Shafie and Klippel 1981). They note that the most commonly associated anomalies are gastro-intestinal, including imperforate anus.

Also associated with oesophageal atresia is prematurity (Warkany 1971), Cywes et al (1976) note that 33% of babies in their study were premature.

(d) Incidence

Cywes et al (1976) believe that the anomaly is more common than the estimated incidence because many infants die of pneumonia before the correct diagnosis is made. This anomaly is fairly common and estimated at 1:3 000 (De Wet 1984)

(e) Presenting signs and symptoms

One of the first signs is maternal hydramnios (excess of amniotic fluid) as the foetus cannot absorb this in the normal way because of the anomaly (Cywes et al 1976). As far as the infant is concerned excessive frothy, foamy salivation is the most obvious symptom as the saliva cannot reach the stomach by the normal pathway. Saliva accumulates in the upper pouch and spills over into the trachea. The child appears hungry and eager to feed, but any attempts at feeding are followed by gagging, coughing and respiratory distress, and if continued the child may die from aspiration of food and pneumonia. The diagnosis will be aided by passing a catheter down the oesophagus as it will be unable to be pushed any further in the presence of the obstruction. The diagnosis is confirmed radiologically.

(f) Treatment

Oesophageal atresia is a neonatal emergency and requires immediate treatment. Treatment, i.e. type of operation is determined by the infant's birth weight and the presence of associated anomalies. It involves surgical repair - either a primary or staged operation. Primary repair which is used for full-term, healthy infants involves tying off the fistula and later surgery on the oesophagus when the infant is more healthy. If the gap between the two blind ends of the oesophagus is too wide, a piece of colon is used to join the two ends. Post-operative complications include pulmonary infections, recurrent trachea-oesophageal fistula and anastomotic leaks (Cywes et al 1976; Cudmore 1978). There is also a high risk of pneumonia.

(g) Results

Cudmore (1978) notes that comparison over the years is not strictly reliable because of changing factors contributing to improved survival.

The results depend on the condition of the baby on admission and the associated anomalies, but the survival rate in full-term babies according to Cywes et al (1976), should be about 90% if they do not have other major anomalies or severe pneumonia. Cudmore (1978) notes that the quality of life is poor only in a minute proportion of babies born with this anomaly.

A study of 86 patients seen over a 12 year period at The Children's Hospital revealed that the overall survival rate was 65% (Cywes et al 1976). At present the survival rate is much higher.

(h) Residual problems

The most difficult period for children born with this anomaly is the earliest months or years. This early period is very stressful for parents, and in the earlier years when the respondents in the present study underwent surgery, they often had to spend long periods of time in hospital. This, coupled with the fact that the mother initially could not feed the child, sometimes for months, placed a strain on early mother/parent infant bonding and could have negative implications for later parent/child relationships.

After the initial period the child should be relatively symptom-free. Long term residual problems may include being underweight, oesophageal stricture, choking, swallowing difficulties, recurrent chest infections and a brassy sounding cough. Overall, one would expect the oesophageal atresia respondent to be completely normal in the long term with no residual problems, or mild problems.

B. Hirschsprung's disease (Congenital intestinal aganglionosis)

Although Harold Hirschsprung was not the first to describe this anomaly, he was the first to recognize the fact that a congenital malformation might be the cause of the disease (Lister and Rickman, 1978).

(a) Pathology

Hirschsprung's disease refers to an absence of ganglion (nerve) cells in part of the large intestine causing an abnormally distended bowel, proximally. Therefore faecal material which is normally propelled through the large intestine towards the rectum and anus by peristalsis, by the intestinal walls, is obstructed at the point of the intestine which has no nerves. This part of the intestine, before the aganglionic part, gets wider and the abdomen distends. Either a short segment or a long segment can be affected and in some cases the whole large intestine is affected, ie total colonic aganglionosis (Holder and Ashcraft 1980).

(b) Aetiology

There is a definite hereditary aspect, although De Wet (1984) notes that the role has remained unclear and that environmental factors also seem important. Sieber (1986) notes that in 3-5% of cases a genetic or familial factor can be implicated.

(c) Associated anomalies

Shafie and Klippel (1981) point out that the frequency of associated anomalies in infants with Hirschsprung's disease is low compared to the incidence found in other anomalies of the gastro-intestinal tract. They do point out, as do many authors, that there is an association

with Down's Syndrome.

(d) Incidence

The exact incidence is not known. Estimates range from 1:1000 to 1:20 000 - 1:30 000 (Lister and Rickham 1978) with the latest estimate at 1:5 000 (Sieber 1986). The incidence in Cape Town is 1:2000 (Louw 1978). This anomaly is usually more common in males than females and Warkany (1971) notes that the ratio of females to males is 1:4,5. Short segment Hirschsprung's disease is more common in males. The longer the segment the more equal the distribution among the sexes.

(e) Presenting signs and symptoms

The first sign is the delayed passage of meconium with subsequent constipation (Sieber 1986). In the typical patient, difficulties in bowel movements begin in the first weeks or months of life and subsequently the colon becomes enormously distended and filled with gas or faecal material leading to abdominal distension, poor nutrition and respiratory distress (Warkany 1971). Other symptoms include bile-stained vomiting and reluctance to feed. When a rectal examination is done, there is often an explosive passage of flatus and meconium or faeces. A positive diagnosis is made by a histological study from a rectal biopsy substantiated by a barium enema and rectal pressure study. An examination of the bowel shows no ganglion cells in the affected segment of bowel if the diagnosis is positive (Lister and Rickham, 1978).

(f) Treatment

If Hirschsprung's disease is left untreated, there is a very high

mortality (40%) in the first year of life (De Wet 1984). Initially obstruction and distension can be relieved through bowel washouts, but there is the danger of recurrent obstruction and necrotizing enterocolitis. Surgery is not performed immediately and a colostomy is initially performed. Between 6-12 months a bowel resection and pull-through operation is performed and the colostomy closed. Mortality in the neonatal period is high (Lister and Rickham 1978) and post-operative complications include leakage from the anastomosis and enterocolitis.

(g) Results

Louw and Cywes (1967) report on the overall results of 80 patients treated at the Children's Hospital for Hirschsprung's disease during the 1952-1965 period. They found that 66 (83%) had good final results. These results have improved with greater knowledge and surgical skills.

(h) Residual problems

These vary from patient to patient, but can include bladder and genital problems, problems with incontinence (especially at night), constipation, obstruction, and distension (from flatus). The residual problems in the early years and childhood period are frequently similar to the residual symptoms associated with high anorectal malformations, i.e bowel problems. Potentially problematic ages are the toilet training stage where parents attempt to get the child to establish a normal stooling pattern and primary school where continuing bowel problems could cause ostracism by peers. These problems usually diminish during adolescence and these patients should be relatively normal after this. They might have occasional problems

with flatus, diarrhoea and/or smearing. In adulthood another problem could emerge - that of fertility. Some respondents have problems with fertility and possible causes could include, especially in females, the adhesions and scarring associated with repeated internal surgery. Problems with fertility could have negative psycho-social consequences.

Overall one would expect the Hirschsprung's disease respondents to have some residual problems which would gradually diminish with age.

C. Anorectal malformations (includes discussion of all anorectal malformations, although only high anomalies were used in the study).

In the 7th century Aeginetta described a surgical procedure for anal obstruction, although a more precise surgical procedure was performed in 1787 by Bell (Templeton and O'Neill 1966).

(a) Pathology

Anorectal malformations refer to an abnormality of the anus and/or rectum. More than 30 different forms of anorectal malformation have been described, ranging from minor abnormalities of the anus to complicated deviations of the anus and rectum (Cook 1978). These anomalies are divided into high, intermediate and low, according to the termination of bowel in relation to the muscular floor of the pelvis. In a series of 287 patients treated at the Children's Hospital from 1954-1969, 40% had high anomalies, 40% low anomalies and 15% intermediate anomalies (Louw et al 1971). Malformations are also classified according to the patient's sex. Warkany (1971) notes that approximately 70% of patients have fistulas connecting the rectum with the perineum or urogenital tract. In the high anomalies there are defects in the muscular sphincters. Jones and Woodward (1986) point out that the

internal anal sphincter is always absent in high anomalies but the external sphincter varies from normal to barely functioning.

(b) Aetiology

The aetiology is not known except that there is an arrest or abnormality in the embryological development of the anus and rectum (De Wet 1984). Although some familial cases have been reported, they appear randomly.

(c) Associated anomalies

Urogenital malformations are the most common associated malformations, but oesophageal atresia may also occur (Shafie and Klippell 1981). The former are usually serious in those with high anomalies. Other associated anomalies are vertebral, skeletal, heart and alimentary tract anomalies (Warkany 1971).

(d) Incidence

This varies from 1:18 000 births in Cape Town, South Africa (Louw et al 1971), to 1: 10 000 births in France (Cook 1978). Louw et al (1971) found the highest incidence in the Coloured and White population groups, while amongst Blacks the incidence was lower. The reason for these differences is not clear. A recent estimate puts the incidence at 1:5 000 (Templeton and O'Neill 1986). There is a slight preponderance of males, especially those with high anomalies.

(e) Presenting signs and symptoms

The problem should be noticed immediately after birth by simple inspection, but it is often missed. The anus is not present in the normal place and further examination may detect an abnormally positioned opening, eg. perineum, vagina or urethra through which meconium passes.

(f) Treatment

The type of anomaly will determine the nature and complexity of the surgical procedure required. High anomalies are always serious. Complications include severe obstruction, abnormalities of the vertebral and urinary tract and abnormal pelvic muscles. Initially these cases are treated with a colostomy to relieve obstruction and at 6-12 months of age reconstructive surgery is commenced. The colostomy is usually closed at a later stage. Post-operative management includes anal dilatation before the colostomy is closed to prevent stenosis (Davies et al 1979). Dilatations must be continued by parents to prevent stenosis of the anus.

(g) Results

Cywes et al (1971) noted that the results were disappointing in terms of absolute continence. This has changed recently as surgery has improved. There is a higher mortality in high anomalies and more complications post-operatively (Cook 1978). At the Children's Hospital this figure for high anomalies was 25%; in 16% of cases death was due to associated anomalies and 6% to delay in diagnosis (Cywes et al 1971). As far as long term results are concerned, in an assessment of 66 patients with high anomalies three years after the operation, 41% had good, 35% fair and 24% poor results, mainly based on the degree of faecal continence (Cywes et al 1971).

(h) Residual problems

The main problem is incontinence and in high anomalies this is frequently a long-term problem. Problems often arise between the child's third and fifth year when toilet training commences. As De Wet (1984) points out, a child who has never been continent before, may not see the advantages of continence and may have reduced motivation to learn how to control his/her

bowels. When the respondents in the present study underwent surgery their anus resembled a stoma and the mucous on the edge could cause mucosal and faecal smearing in the respondent's pants. Lack of sphincter control causes incontinence as respondents cannot control their bowels and sometimes cannot feel when they need to defaecate. Furthermore this lack of sphincters also means that some respondents cannot control flatulence. Their flatus is also especially foul smelling. These respondents also often had their colostomy for quite a long time in those days, sometimes several years.

Other problems associated with anorectal malformations are urinary and genital problems. Urinary problems could be due to associated urological anomalies or could be caused by the type of surgery necessary to correct the anomaly, i.e. can involve disturbing nerves associated with organs involved in urination. This surgery could also cause sexual dysfunctions. Interference with nerves, scarring and adhesions could also cause infertility in males and females.

In the child's early years, the parents are mainly involved in handling the child's bowel and/or urinary problems, dilatating the child's anus, and in toilet training. This can be a very difficult time for parents and can interfere with the parent-child relationship (De Wet 1984). Should incontinence continue, the child may experience problems at school, eg. not being able to participate in sport, absences from school, having 'accidents' at school (which could cause peer rejection and ridicule). Wearing a colostomy bag could also have caused similar problems during adolescence and adulthood. Anticipated problems, resulting from incontinence and sexual difficulties, would include interpersonal problems and, if fertility is a problem, possible marital difficulties. Overall, one would expect the high anorectal malformation patient to have many residual problems in the long term.

2.3. CONCLUSION

The oesophageal atresia (OA) respondents are expected to have no to mild disability/residual problems, the Hirschsprung's disease (HD) respondents are expected to have moderate disability/residual problems and the high anorectal malformation (AM) respondents are expected to have severe disability/residual problems.

CHAPTER 3

THE PSYCHO-SOCIAL EFFECTS OF ILLNESS/DISABILITY - A LITERATURE REVIEW

Prior to conducting research into a particular topic, it is important to review the available literature to assess what research has been conducted in the past and to obtain direction for future research.

A literature search, conducted by the Institute for Biomedical Communication (Medical Research Council), revealed a paucity of literature concerning the psycho-social effects of illness/disability in adulthood. Studies have tended to focus on children and to a lesser extent on adolescents.

For this reason, and because a person's childhood and adolescence will influence his/her adulthood, childhood and adolescent studies are examined prior to adult studies.

There is very little research concerning the anomalies under discussion. Therefore the scope of the literature review has been widened to include studies on the psycho-social effects of illness/disability in general and studies on some congenital anomalies which were seen as similar to the ones under discussion. Referring to the latter, certain spina bifida studies were examined, as this anomaly is seen to be comparable in some ways to Hirschsprung's disease and anorectal malformations as spina bifida is a congenital defect, requires surgery and is often associated with bowel and/or urinary problems. Cleft lip/palate is in some ways comparable to oesophageal atresia in that it is a congenital defect, frequently requires childhood surgery and involves early feeding difficulties (although it is a more obvious physical defect).

Therefore this chapter looks at:

3.1 Childhood studies

- A. Illness/disability studies in general
- B. Studies on the anomalies under discussion

3.2 Adolescent studies

- A. Illness/disability studies in general
- B. Studies on the anomalies under discussion

3.3 Adulthood studies

- A. Illness/disability studies in general

3.4 Conclusion

3.1 CHILDHOOD STUDIES

A. Illness/disability studies in general:

It has been suggested that physical illness or handicap in childhood is associated with a significant risk of psychological sequelae (Mattsson 1972). Various studies note the negative psycho-social effects on children with chronic illness or congenital defects (Sandberg 1976; O'Moore 1980; Pless and Roghmann 1971; Green and Solnit 1964). The latter coined the term 'vulnerable child syndrome' to describe children born with congenital anomalies who present with certain common symptoms including pathological separation difficulties, infantile behaviour, bodily over-concerns and school under-achievement. However, there is controversy about both the existence and the nature of these relationships (Stein and Jessop 1984) with some evidence to the contrary (Tavormina et al 1976). Sandberg (1976) points out that Anderson's (1976) study showed no significant differences between disabled children and controls and Mattsson's (1972) review article on adaptation to long-term physical

illness notes that many studies report a surprisingly adequate psycho-social adaptation of children followed to young adulthood.

Various studies have examined the different factors that influence the psycho-social effects of physical illness or disability on children. Factors include the severity of the disability, the type of disability and the family's reaction to the handicapped member. The latter is important for the present study which involves examining the effect of the severity of residual problems/disability on psycho-social functioning. According to Mattsson (1972) the literature indicates that the nature of a specific illness has less influence on a child's successful adaptation than such factors as his developmental level, available coping techniques, the quality of the parent-child relationship and the family's acceptance of the handicapped member. Heller et al (1985) note that results of studies implicating severity, type of disorder and family reaction to disorder are inconclusive and they therefore conducted a recent study on birth defects to examine these factors. They found that the type of disorder and its severity are associated with remaining or becoming maladjusted. They did not find family functioning to be significantly related to maladjustment. The stresses of chronic physical illness on children are well documented in the literature (Lowit 1973). These can include the traumatic effects of hospitalization, separation from parents and surgery, which have been discussed (Chapter 1).

Referring specifically to surgery, Schultz et al (1983) note that many studies document the negative emotional effects that occur after surgery, including anxiety, fears and regression. Blotcky and Grossman (1978) compared childhood genito-urinary surgery (which can

be compared to surgery for anorectal malformations) and ENT surgery. They found a significant association between childhood genito-urinary surgery and emotional disturbance but no similar relationship between ENT surgery and emotional disturbance. It seems that the effects of surgery may be related to the part of the body which is operated on. This is important to bear in mind when examining the anomalies under discussion, ie the effect of the particular part of the body being operated on.

A study on latency-aged children with spina bifida by Johnson (1984) shows that a delay in reaching important physical milestones, eg continence, hinders the achievement of standard psycho-social developmental goals, but it does not exclude their being achieved.

A study by Kapp-Simon (1986) on 50 children, aged 5 to 9 with cleft palate, cleft lip or both, compared to a control group by means of a pictorial questionnaire, showed that cleft lip/palate children had a lower self-concept, were less socially adept and were more frequently sad and angry compared to peers.

The stress of the chronic illness or disability is an ongoing one for the child and his family. It is important to consider the developmental stages that the child is going through in order to fully understand the effects which surgery and the illness may have. Schultz et al (1983) note that the timing of surgery is very important and they suggest that it can influence the psychological effects. They point out that the literature indicates that there is a different response to surgery at different ages.

As already discussed, children with oesophageal atresia have feeding problems at a stage of development where feeding is a vitally important task. Similarly, children with Hirschprung's disease and anorectal malformations have problems with bowel functioning at a stage of development where toilet training is an important task to master. The consequences of surgery and/or disability-related problems at these early stages of a child's development may be serious. Only a few studies have been conducted in the field of these anomalies and will be referred to below.

B. Studies on the anomalies under discussion

Referring to oesophageal atresia, a few studies have been conducted. Of the studies and theories which have been proposed and conducted concerning the importance of feeding, one of the most challenging, according to Engel and Reichsman (1979), is Freud's theory that the infant's experience with feeding is a crucial factor in personality development and integration.

In their case study of 9 infants with feeding difficulties, Illingworth and Lister (1964) suggested that there is a long-term effect associated with oesophageal atresia. This group included 3 children with surgically corrected oesophageal atresia who had been fed by gastrostomy for long periods prior to surgery. The children presented with various feeding problems including refusal of solid foods, failure to chew and vomiting. The authors relate these difficulties to a sensitive period which is the optimum age for the learning of each particular skill (eg. chewing solid foods), after which it becomes increasingly difficult to learn appropriate behaviour.

If children are not given solid food to chew at a time when they are first able to chew, they may experience difficulties in feeding.

Later a 'critical period' is reached where the specific pattern of behaviour can no longer be learned. They do point out that the average age for this specific behaviour, eg. chewing, varies in children according to maturation and aptitude. They suggest that in oesophageal atresia the sensitive period may have been missed.

Gibson (1965) studied the effects of trauma in early infancy and later personality development and included 9 patients with oesophageal atresia in his sample of 29 patients who required childhood surgery. He compared the psychodynamic patterns of personality of children born with congenital obstructions to normal children (both groups aged 5 to 8 years) by means of 4 projective techniques - Children's Apperception Test, Draw-a-Person test, Wishes and Fears Inventory and Blacky Pictures. The mothers were given paper and pencil inventories. The results suggest that there were no significantly different psychodynamic personality patterns and emotional disturbances in the oesophageal atresia group. This finding was not expected if one takes into account the hospitalization experiences and feeding difficulties, ie. gastrostomy. However, the authors do note that the sample may be biased as mothers who were well adjusted and/or whose children were well adjusted, may be more willing to participate in research.

Dowling (1977) undertook an intensive case study involving observation of 7 infants with oesophageal atresia and suggests that developmental disturbances found in the infants are related to the absence of activity patterns and incorporative patterns which are regular parts of normal oral organization, ie feeding. Five of the children presented with various difficulties including slow development, lack of forcefulness in their development, low motivation in mastering new activities, not extending learned activities to new situations and regression if difficulties were encountered. Their behaviour seemed to lack motivation and pleasure with the environment. Although they showed preference for their parents compared to strangers at the age of 5 months, this response was also unenthusiastic.

Dowling emphasizes the importance of the oral experience as the earliest organizer of instinctual mental life. He emphasizes the important function of feeding apart from other factors such as mothering and states " ... infants who received no oral feeding or normal oral feeding and who did not experience a regular rhythm of hunger and satiation, but who did receive adequate maternal care apart from the feeding situation, suffered massive dramatic disturbances of gross motor development." The infants failed to develop normal goal-directed behaviour and involvement with both their physical and social environments. He notes that disturbances of motor, perceptual and affective development occur which are related to the failure of normal feeding experience. He notes that even at 11 years, although motor and feeding difficulties were no longer apparent, there were disturbances of learning and relationship difficulties.

Dowling suggests that the physical act of oral feeding provides an important stimulus and organising force for a variety of developmental accomplishments. He feels that hunger is an important motivational force which is extended to other behaviour and in these infants was hindered or non-existent. However, De Wet (1984) points out that this study was limited to early infancy and therefore precludes findings on whether these symptoms remain.

Engel and Reichsman first reported on an intensive longitudinal case study of a child with oesophageal atresia in 1956 (Dowling 1977). They report on a patient, Monica, studied and filmed by a research team over a 25 year period. Dowling (1977) notes that a 1967 report by Engel placed emphasis on the abnormal feeding situation as an important factor in Monica's abnormal development.

An important theme in the study of Monica was that the origin of object relationships is an assimilative process which is organized orally (Engel and Reichsman 1979). They report similar findings to Dowling's (1977) study with Monica showing problems of gross motor co-ordination, little motivation or initiative and a tendency towards imitative behaviour.

De Wet (1984) comments on a study by Dera et al (1980) of the psychological development of children (3 to 6 years) who underwent surgery for congenital atresias, included 10 children with oesophageal atresia. A relationship inventory was administered and disturbances of psycho-social development were

found, eg. disturbances of contact, separation anxiety and regressive behaviours in those with more severe malformations or in whom the operative repair was functionally incomplete. IQ scores were within the normal range. The most notable result was a lack of aggression in all the groups studied. The authors point out that the frequency and duration of early separation of mother and infant, together with the frustrated feeding relationship, led to the findings.

Lindahl (1984) studied the long-term prognosis of 43 successfully operated oesophageal atresia children with emphasis on physical and psychological development. The group ranged in age from 10 to late teens and was compared to a control group aged 8 to 12 years. The Weschler Intelligence Scale for Children (WISC), the Human Figure drawing and the Rorschach tests were administered to the patients and a control group. In examining intelligence and body image the psychological development of these children appears normal and the authors describe the patients as a group as intelligent, introversive and somewhat inhibited. They conclude that for successfully repaired oesophageal atresia, '... the long-term prognosis is good, physical and psychological development seems normal and the quality of life is excellent for the majority of patients' (Lindahl 1984:10).

Koop et al (1975) studied the social, psychological and economic problems of the patient's family after successful repair of oesophageal atresia. Thirty-one families whose children had been operated on from 10 to 25 years previously were studied.

They found that while most parents admitted some problems in the past, eg feeling socially isolated, they seldom admitted problems in the present. The greatest difficulty occurred in those children who formed a very close relationship with their mother. Most parents thought their child's general disposition was happy and only children whose difficulty in swallowing continued, seemed to be aware of their problems. In reaction to frustration, girls responded with sulky behaviour and boys with anger. The only embarrassing residual symptom according to the parents was the brassy cough some patients had and some difficulties in swallowing.

De Wet's (1984) study appears to be the only South African study examining psycho-social effects of the anomaly groups under discussion. She focuses on the psycho-social effects of these anomalies on the families of patients (these reactions would in turn influence the patient). By means of in-depth interviewing, she studied 83 families with 90 affected children with oesophageal atresia, Hirschsprung's disease, anorectal malformations and cystic fibrosis. (The latter will be excluded from the discussion). She included 23 cases of oesophageal atresia, where the patient had undergone surgery at the Red Cross War Memorial Children's Hospital. She found that 96% of parents were very anxious about the possibility that their child would die. It seemed that there were fewer effects on the family relationships from the illness than for other anomaly groups, with a lower incidence of strained marriages in families with the children who had oesophageal atresia. She found that while there were a few problems with the patients in their early years, eg.

choking on foodstuffs, chest problems, there were very few problems after the age of approximately 8 years and no problems relating to school (academic or social problems).

The above studies reveal no clear indications of psycho-social maladjustment associated with oesophageal atresia. The most recent research, ie. Lindahl (1984) indicate a good psycho-social prognosis for these children. While the research on oesophageal atresia has frequently been intensive and in depth (and herein lies its value), most of the research is outdated, conducted on very small samples, lacks controls and, with the exception of Engel and Reichman's and De Wet's study, has examined children at a very young age. Research involving long term follow-up is clearly needed.

Referring to Hirschsprung's disease, De Wet (1984) notes that there is no specific literature dealing with this illness, although Tejani et al (1978) include patients with this diagnosis in their study concerning the effects of gut surgery. They found that neonates who had undergone gut resection are at a higher risk for delay in height and weight gain compared to those who have gut surgery without resection. They also found some perceptual motor defects on the Bender test, although they had not excluded the possibility of this being due to neurological dysfunction.

Valman (1974) used the Draw-a-Person test and school reports to study the intelligence of 8 children (aged 3 to 14 years) who had had neonatal resection of the ileum. They were compared with a

group of healthy controls and a group of patients with cystic fibrosis. He found no significant differences between the groups regarding intelligence.

In the previously mentioned study by De Wet (1984), 30 patients with Hirschprung's disease were included. Initial problems experienced by parents included a delay in diagnosis and separation from their infant. She notes that extreme difficulty was experienced by 8 families in obtaining a diagnosis, and these families revealed feelings of lasting reproach and hostility at consultants concerning this delay. Other problems included not having the operation properly explained to them, fears about losing the child and difficulties with staff. There were many unpleasant residual problems that parents had to deal with, including handling of colostomies. Toilet training was found to be very difficult and led to many family arguments. These problems affected the parent/child relationship to varying extents. As regards family relationships, 43% of families had strained marriages which were often attributed to the anomaly.

There was a high incidence of problems for these patients once they began attending school. Seventy-six percent of those attending school had experienced unpleasant incidents at school, eg staining, smelliness, flatulence, diarrhoea. Many parents remained unaware of how patients had coped even though they found evidence of incontinence. There seemed to be a lack of communication between parents and children concerning problems due to their embarrassing nature. Colostomies were a great problem, being difficult to hide and handle effectively. Academic

problems were experienced by 29% (6) although only 1 of these patients had low intelligence. Regarding sport, 52% (11) were actually involved in at least one type of sport, but many had given up because of fear of close contact and lack of privacy when changing into sports clothes. They also experienced peer ridicule and ostracism which has been described by researchers of other anomalies, eg spina bifida (Dorner 1976).

There is a great need for research in this area as the limited work that has been done suggests that children with Hirschsprung's disease face a range of psycho-social and physical problems.

Referring to children with anorectal malformations, Mies et al (1978) note that research on children with congenital atresias, including anorectal malformations, has largely been ignored. They also note that early separation and the first reaction of parents, especially the mother, seems to have a great influence on parent/child interaction and on the child's emotional development.

In the previously mentioned study by Gibson (1965), 5 patients with imperforate anus were included. It was found that children with problems relating to elimination and the need for post-surgical manipulation of the anus by the mother showed many signs of disturbance, and the mothers showed deviant attitudes towards their child.

In the study by Dera et al (1980) (previously discussed), 20 children with anal atresias were included (aged 3 to 6 years). They demonstrated some separation anxiety and disturbance of contact. De Wet (1984) notes that it was found in Dera et al's study that intellectual development showed no meaningful impairment, but nursery school and school were often delayed because of faecal incontinence. Only 8 of the patients were fully continent. Interestingly, she points out that in some cases incontinence was not felt to be only of functional origin, but related to painful anal dilatations which upset the mother/child relationship. She also points out that the handling of colostomies detracted from the pleasurable aspects of mother/infant interaction, which in some cases led to rejection by the mother. An unexpected finding showed retarded speech development in anal atresia patients compared to the other anomaly groups. The authors suggest that prolonged hospitalization could not have caused this problem as patients with oesophageal atresia had the longest hospitalization. They tentatively suggest that anal dilatations were a contributory factor, which is supported by other studies.

In De Wet's (1984) study 20 patients were included with anorectal malformations. Difficulties experienced by parents included delay in diagnosis, difficulties with staff in the maternity home and inadequate viewing of the baby before being transferred to the hospital, ie. separation. The diagnosis was experienced as a crisis by 65% of the parents because they feared for their child's life. Residual symptoms were difficult for parents to handle and similar results to Hirschsprung's disease were found

with problems with colostomies, anal dilatations and toilet training. She notes that this anomaly group showed the most problems with residual symptoms in the long-term adaptational phase. There was interference with family relationships with 50% of marriages being strained. As regards school, a large percentage (70%) attending school experienced embarrassing incidents at school. It was found that when fathers had not been closely involved in the handling of symptoms, boys did not discuss difficulties experienced at school. A similar percentage to the Hirschsprung's disease anomaly group participated in sport (50%). It appears from De Wet's study that the anorectal malformation patients are slightly more at risk for psycho-social maladjustment than Hirschsprung's disease patients.

In a study of 120 patients with high imperforate anus (Ditesheim et al 1987), the relationship between quality of life and faecal continence was examined. They found that in the pre-school age child who had faecal incontinence, having strong family and social support systems can be enough to protect the child and ensure he has good quality of life. An understanding teacher at the age when the child enters school combined with various strategies to avoid accidents can protect the child from social embarrassment and ostracism. Strategies include dietary management, medication, toilet training and enema programmes.

Again the psycho-social problems that were found in this limited research warrant further investigation in this field, especially in the sphere of long term follow-up studies.

The majority of these studies have referred to the effects of

these anomalies on young children. Yet one would expect that children entering school would be dealing with other equally distressing difficulties in the case of the Hirschsprung's disease and anorectal malformation anomaly groups. At this stage, described by Freud as the 'latency stage', achievement and conformity are key issues. It is possible that embarrassing residual symptoms at this age and under-achievement either academically (because of long absences) or on the sportsfield (because of physical problems) may interfere with the child's body image, confidence and self-esteem.

The studies above reveal that children with AM and HD are at risk for psycho-social problems.

Although there is a lack of consensus concerning the psycho-social effects of chronic physical illness or disability in childhood, the research which has been conducted seems to indicate that the children in the anomaly groups under discussion may be at risk for psycho-social problems.

3.2 ADOLESCENT STUDIES

Adolescence is a difficult time for most people in that it involves dealing with issues like sexuality, identity-seeking, physical changes and dependency/independency conflicts. The teenager who has difficulties like a chronic, physical illness or disability may find their difficulties heightened at a time when self-esteem, independence and conformity are very important. It would be expected that some teenagers may experience self-esteem problems, problems with independence (especially if physically dependent on parents when ill, or if parents are over-protective), problems

with relationships and sexual problems.

A. Illness/disability studies in general

While there is a general lack of research conducted on the chronically ill or disabled adolescent, some studies over the past decade have focused on the psycho-social functioning of the adolescent with a chronic illness or disability (Tavormina et al 1976; Orr et al 1984; Kellerman et al 1980). There is also a controversy over the psycho-social effects of chronic illness and disability with some studies reporting maladjustment (Mattsson 1972; Hayden et al 1979) and others focusing on positive aspects like the resilience and coping skills of adolescents (Tavormina et al 1976). A South African study (Straker and Kuttner 1980) revealed no psychological differences between adolescents with a chronic illness (cystic fibrosis) and a healthy control group (cystic fibrosis is in some ways a comparable congenital anomaly to anorectal malformations and Hirschsprung's disease in that it also has anti-social symptoms like smelliness). Regarding different factors which influence psycho-social adjustment, Castree and Walker (1981) found that it was self image which had the most influence on the social isolation of spina bifida patients, not the severity of the disability. In fact, Hayden et al (1979) found that some of his subjects who were least disabled viewed themselves as very unhealthy whereas some of them who were very disabled reported no physical problems. Findings suggest that the ability to cope with physical and emotional situations is not determined by the severity of the disability, but is determined by all the environmental support systems since birth.

It is useful to examine studies concerning spina bifida, for the reasons discussed previously, and because this disability has been the focus of research on adolescent physical disabilities. Despite the obvious physical difficulties, problems that have been identified in adolescents with spina bifida include self-esteem problems (Blum 1983; Hayden et al 1979; McAndrew 1979), social isolation and adjustment problems (Dorner 1976; Blum 1983; Hayden et al 1979; McAndrew 1979), employment problems (Lorber et al 1973) Dorner 1976; Blum 1983; McAndrew 1979), sexual worries and problems (Hayden et al 1979); Blum (1983) depression (Dorner 1979; McAndrew 1979) and social embarrassment over physical problems (McAndrew 1979). Wabreck (1986) notes that embarrassment from bladder or bowel accidents affects psycho-social adjustment negatively, especially self-esteem. Farrow (1980) notes that male adolescents with genital abnormalities suffer great problems in psycho-sexual adjustment.

Another comparable physical handicap (for reasons previously discussed) which has been addressed in the literature concerning adolescents is cleft lip and/or palate. Again there is controversy over whether this handicap leads to maladjustment or not, but it seems that no clear or significant relationship exists between the handicap and poor adjustment (Starr 1978; Richman 1983). Starr's (1978) findings support the notion that a physical anomaly per se is not a factor in explaining social behaviour. He also notes that it is the patient's perception of his disability that is associated with whether adjustment is poor or not. Furthermore Richman et al (1985) note that cleft lip and palate adolescents with good adjustment had realistic perceptions of appearance, while the poorly adjusted group had unrealistic perceptions of their appearance and behaviour.

B. Studies on the anomalies under discussion

In Ditesheim et al's (1987) study on children with high imperforate anus, they note that children who have not achieved faecal continence will start experiencing problems when they reach adolescence and that quality of life may diminish considerably. They feel that at this stage family and social support systems are not enough to cover up the problems, eg. accidents and odour. Furthermore, they note that some families become less supportive as the child's physical and social adjustment problems carry on or get worse.

There appears to be little literature relating to adolescents concerning the anomaly groups under discussion. While one can speculate on the difficulties that would be encountered when children with embarrassing residual symptoms reach adolescence, further research is needed to highlight the issues involved.

3.3 ADULTHOOD STUDIES

It is during the adult years that the effect of a chronic illness or disability can often be better assessed, as the patient's personality is better formed and generally more stable than before.

The life tasks usually being dealt with at this stage, ie young adulthood, include choosing a partner, establishing a career, starting a family, setting up a home and so on. It is within this context that the psycho-social adjustment of young adults in the various anomaly groups must be viewed. It would be expected that patients who still had urinary and/or bowel and/or sexual problems, may be experiencing difficulty at this stage of development.

A. Illness/disability studies in general

As there are no studies dealing specifically with the psycho-social effects of the various anomalies in adulthood, it has been necessary to draw on general studies concerning chronic illness or handicap and specific studies concerning handicaps that have some similarities with the anomalies under discussion.

Harris (1982) examined the symptomatology of abnormal appearance in 54 patients ranging from childhood to adulthood by analysing written accounts by patients. While this study concerns overt disabilities, some of the findings are probably comparable to other disability groups. Among defence mechanisms used were restrictions of various aspects of their lifestyles to avoid those situations which they had learnt from experience to be embarrassing, secrecy about the defect and trying to camouflage or hide it in some way. Two-thirds of the patients described feelings of inferiority, unattractiveness, lack of self-confidence and insecurity.

Barron (1985) in a study of 30 male adult patients between 17 and 44 used the Weschler-Bellevue Intelligence Scale, Rorschach Test and a Sentence Completion Test to compare patients with seen (eg. poliomyelitis) and unseen (tuberculosis) physical handicaps. Results suggest no significant difference between the two groups, rather there was a similarity of trends. This study is interesting in that the anomalies under discussion are a largely unseen handicap, except in intimate situations.

Referring to cleft lip/palate research, Heller et al (1981) studied 96 young adults (between 18 and 27 years) born with a cleft lip/palate to evaluate their psycho-social adjustment using telephone interviews.

Psycho-social functioning was judged to be adequate for 67%, marginal for 23% and clearly inadequate for 10%. Difficulties relating to the handicap included problems at school (25%), and past problems with social life (over 50%), although only 6% admitted to problems with the opposite sex. Some experienced problems with their social life (56%), had few friends (24%) and few leisure activities (nearly 50%). Whilst 54% of the 80 with jobs were in professional or white collar employment, nearly 30% reported having difficulties finding employment and less than 45% had job satisfaction. Other studies also report fewer friendships, decreased participation in social activities and lower marriage rate (Peter and Chinsky 1974).

Peter and Chinsky (1974;1975), studied 196 patients with cleft palate or cleft lip and palate aged 24 to 54 and compared them with a group of siblings in the same age group and a control group. They were assessed on marriage and vocational/economic aspects by means of a self-administered questionnaire. These patients were found to marry at a significantly lower rate, marry when older, have fewer children per years married and have more childless marriages. Although cleft lip/palate adults functioned within normal limits with regard to employment, their levels of income were lower than other groups and overall the authors conclude that cleft lip/palate adults experience some limitation as regards vocational and economic aspects of their lives.

In Clifford et al's (1972) study of 98 adults with cleft lip-palates, in the 20 to 30 age group, the level of satisfaction with themselves, their bodies, their appearance and the treatments they received was high. They perceived their clefts as having had relatively little

influence on their lives.

Laurence and Beresford (1975) studied 51 adults with spina bifida aged between 17 and 56 years. While only 17 were completely continent the authors note that this did not seem to impede social relationships and that the most important factors in making friendships in childhood are school experiences, the personality of the patient and mobility. They did however note that spina bifida seems to reduce the likelihood of marriage.

Evans et al (1974) studied 202 spina bifida adults and found that 93 of the patients were living normal lives, but 109 had serious disabilities. Of the latter group who had serious locomotor problems and/or incontinence, 58 work regularly, 31 had never worked and 24 had married.

Dunne and Shurtleff (1986) studied 37 young adults aged 19-27 and reported that 25% of their patients were employed and 48% of females and 23% of males were married. They found 10% to have low self-esteem and 14% had no close friends. They point out that their previous study (1975) revealed that one-third of adults over 19 years of age had psycho-social problems severe enough to cause failure to meet expected goals.

Another congenital anomaly group which may be comparable to anorectal malformations are patients born with hypospadias (a malformation of the urethra) as this anomaly requires urinary surgery. (To correct some types of anorectal anomalies, surgery involves the urethra).

Schultz et al (1983) present a comprehensive review of the literature regarding the effects of surgery, more specifically hypospadias surgery on psychological development. They note that Erikson and Von Hedenberg (1971) found that men who had had hypospadias repair tended to abstain from sexual intercourse. Berg et al (1981) in a study of 34 men operated on for hypospadias in childhood and a control group interviewed subjects concerning their sexual and social adjustment which was virtually the same in the two groups. They did, however, have later first sexual experiences and less frequent sexual activity than controls. The hypospadiac patients were found to be in less qualified professions than controls, despite similar intellectual abilities.

Similar results were found in Farkas and Hynie's (1970) study of 130 patients, 18 and over, with repaired hypospadias. They note that even though there was adequate repair and most patients could function normally more than half of them abstained from sexual intercourse, because of feelings of abnormality and inferiority.

Berg and Berg (1983) studied 33 adult hypospadias (aged 20 to 35) and compared them to matched controls by means of a Rorschach Test in a double blind design. The hypospadiac patients differed significantly from the controls in showing more neurotic traits, lowered self-esteem and capacity for interpersonal relations and higher anxiety and hostility levels. The authors conclude that these results compare well with the psychoanalytic theory on the role of castration anxiety in the formation of neurosis.

However, Schultz et al (1983) point out that Avellan's (1976) study found that hypospadiac patients had normal development and only a slight delay in first sexual experiences.

As regards the different factors which influence the psycho-social effects of illness or disability, in a study on cystic fibrosis by Pinkerton et al (1985), coping patterns in adult life were examined. Substantial differences were found between a coping and non-coping sample. The non-copers compared unfavourably with the copers in 4 main areas - vocational records, level of family support, awareness of anomalies in sexual function and knowledge about their illness. In another study by Berg and Berg (1983) on hypospadias no correlation was found between severity of malformation and personality. Moos et al (1965) note that the greater the functional incapacity in rheumatoid arthritis patients, the more negative the psycho-social symptoms.

Felton et al (1984) studied stress and coping behaviour in chronically ill middle-aged and elderly patients. Although older than the anomaly group under discussion, the results are interesting. They found that coping strategy tends to be minimally explained by medical diagnosis. They suggest that there is a need for more research and some modification of current theories of coping.

3.4 CONCLUSION

It seems that there is a lack of consensus concerning psycho-social effects of illness/disability in adulthood, as was found in the adolescent and childhood studies. However, it does seem that under certain circumstances and in certain anomaly groups, there are

negative psycho-social consequences. It appears from the literature that the anorectal malformation patients and the Hirschsprung's disease patients (to a slightly lesser extent) are at risk for psycho-social problems. While earlier literature indicates that oesophageal atresia patients might be at risk for psycho-social problems, more recent research indicates that the risk appears minimal.

There appears to be minimal research on the anomaly groups under discussion. Furthermore, there appears to be a lack of research in general on young adults. It is clear that research is needed in this area, ie. young adulthood, as adults at this age deal with different life tasks to older adults. Research concerning the anomalies under discussion is thus clearly indicated, and this would contribute to solving the larger problem of limited research in the area of all adult handicaps.

CHAPTER 4RESOURCES AND FACILITIES

There are few medical resources and facilities, and no organizations aimed specifically at patients born with the malformations under discussion, as reflected internationally.

There is a problem in examining resources and facilities for these people, as there is a lack of clarity as to how to classify these groups. Are they chronically ill, or are they physically disabled, or both? Some of them are chronically ill all their lives, and others are completely healthy after their initial malformation has been surgically corrected. Their disability is not really an obvious physical one, except in certain situations where scarring is obvious. However, for some Hirschsprung's disease and most high anorectal malformations patients, their disability, while not obvious visually, is obvious in certain respects in that their bowel problems cause some of them to smell unpleasant. One could classify them as socially handicapped or disabled or perhaps medically handicapped. There is, however, such a range in occurrence and severity of residual symptoms, that these patients do not really fit into any category of illness or disability. These patients need to be judged on an individual basis to determine what organizations and resources in the community are applicable to them. It must be noted that oesophageal atresia (OA) patients did not have as much need for medical services after the early years, as Hirschsprung's disease (HD) and especially anorectal malformations (AM) patients.

4.1 RESOURCES IN THE PAST ie. those that were available for the respondents in the present study.

The services available for respondents born with the anomalies under discussion were mainly hospital based and usually medical services.

A. Medical Services:

The key people in the patient's life, especially in the early years, were the surgeons and the nurses (and stomatherapists for HD and AM). They provided treatment and management and a follow up outpatient service. The OA patients were followed up in the general outpatient clinic while the HD and AM patients were followed up in a specialized anorectal clinic, as there were more of these patients. For those with HD and AM the stomatherapist would have played an important role in helping parents manage their child's colostomy and bowel problems (including anal dilatations for AM patients). The Children's Hospital has had a specialized Stoma Unit since 1976. This unit consists of a nursing sister and an assistant nurse who, in conjunction with paediatric surgeons and urologists, are concerned with the management of patients with stomas (eg. colostomy, ileostomy).

Other resources which might have been used in the community, were local clinics, hospitals or general practitioners. The Red Cross War Memorial Children's Hospital is officially a children's hospital and only admits patients up to the age of 13. When some of the respondents became teenagers they did not return for medical treatment but tended to deal with their problems by themselves and accepted their situation, even if not ideal. Of course, some patients had no further need for medical services.

For those who had continuing problems the options available were to return to The Children's Hospital or to go to other hospitals, eg. Groote Schuur Hospital, or to private doctors. It must be emphasized that there was, and is presently, a large gap in the services for

these patients, ie. teenagers. This means that they were not followed up and this could have caused problems later. This lack of facilities and resources for teenagers highlights a widespread medical health problem in this country. There is a lack of resources for all chronically ill teenagers. These patients have a dilemma - they are too old for their childhood hospital, yet too young for the adult services available at hospitals. It was also probably difficult in the case of HD and AM patients to go to new medical personnel as their medical problems are often embarrassing, and at a very self-conscious age they might have decided not to seek medical help. The surgeons at the hospital do have an open door policy for their former patients, but this is an unwritten policy which many respondents probably did not know about. Furthermore, new staff do not know the patient operated on by their predecessors. The consequences of patients getting lost when they 'hit the gap' as adolescents could be disastrous. Other authors have highlighted this problem and as Bax et al state: 'Little is known about the health needs of physically handicapped young adults after they become too old for the paediatric service' (1988:1153). In their study of young physically handicapped adults, over half the subjects had health problems severe enough to require treatment, yet less than one-third were receiving any form of hospital care.

On approaching adulthood, patients experiencing problems with fertility could make use of the infertility clinic at some of the main hospitals, eg. in Cape Town there are Infertility Clinics at Groote Schuur Hospital and Tygerberg Hospital. Although mainly dealing with patients with colostomies and ileostomies, stoma units, based at main hospitals could possibly be a resource for patients with HD and AM.

Groote Schuur Hospital has an incontinence adviser who could play a similar role. The problem that seems to arise in connection with these stoma units is that they are very involved with patients with permanent colostomies and do not have the time to sort out the incontinence problems of HD and AM patients, which are not always physiological in nature. There does not seem to be anyone or any department at the adult hospitals specially geared for the HD and AM patients, interested in their difficulties or with the knowledge of these anomalies. The responsibility for these adult patients therefore tends to fall on the shoulders of the paediatric surgeons who are very involved with paediatric concerns.

In examining past health care services it must be pointed out that this hospital was not always racially integrated. Treatment for different race groups did not differ. However, resources and facilities in the rural areas differed from urban areas, and still tend to be poorer for 'Black' and 'Coloured' communities.

B. Social Work Services

The Social Work Service has been available to patients since the hospital opened in 1956. However, this service tends to focus mainly on parents and their concerns. Patients who were seen to be having psycho-social concerns tended to be referred to the hospital psychiatrist. After the age of 13 years these patients would have had access to any social worker, psychologist or psychiatric service available to the general population as there was no specialized service for them.

4.2 RESOURCES AT PRESENT ie. those available for the patients presently born with these anomalies.

Services available for these patients are still largely hospital based.

A. Medical Services

The key people in the patient's life in the early years are still the surgeon and the nurses. However, for HD and AM patients there is a full-time stomatherapist and nursing assistant and a specialized Stoma Unit to meet these patients' needs.

Surgery has improved vastly and so have the follow-up services for patients born with congenital anomalies, ie. the Stoma Unit has a special follow-up clinic once a week. They also follow-up their patients to their late teens and personally refer them if they need medical services. In general the same medical services exist for these patients, but they have improved. Although the gap between childhood and adulthood has not been totally bridged, there has been an attempt to extend these services. This Stoma Unit Service i.e. extension of service to late teens combined with the surgeons' open door policy alleviates some of the present problems with service gaps.

As with resources available in the past, present resources in the community include hospitals (especially infertility clinics), and general practitioners.

B. Social Work Services

The Social Work Service has expanded, in the number of available social workers and the scope of their work. However, they still

provide basically the same service, mainly focusing on parents (patients are often initially too young for counselling).

The Stomatherapist also provides advice and counselling and plays a pivotal role in the patient-health care system. De Wet (1984) also comments on the important role the Stomatherapist plays in regard to patients and parents.

4.3 CONCLUSION

It is hoped that as the medical/surgical service has improved so vastly, fewer patients will have long-term residual problems and consequently fewer patients will have the need for social work or other psycho-social services.

CHAPTER 5

RESEARCH METHODS

"Practical issues in psycho-social research fail to receive the attention they deserve, and it is only through a series of unsatisfactory experiences that their importance becomes appreciated." (Yates et al 1983:2376). The present study was fraught with practical difficulties and the following chapter describes the research methodology used within the context of these difficulties.

Watson and Kendall (1983) classify research associated with chronic disease into (1) coping studies which investigate the impact of the chronic disease on the individual, or (2) outcome studies which evaluate the effectiveness of a particular intervention in counteracting the negative impact of the disease. This study concerns the former and the following chapter describes the method used to attempt to assess this impact or effect.

5.1 RESEARCH DESIGN

The design of the research refers to the way in which a researcher will attempt to reach his research goals and how problems encountered in research will be overcome.

Leedy (1985) points out that the nature of the data determines the research methodology. Although the study of the psycho-social impact of illness on individuals is not a new area of research, there is very little research concerning the psycho-social effects of being born with one of the congenital anomalies under discussion. There is also little research concerning long-term effects of illness in adulthood, as highlighted in the review of the literature. Therefore the research design chosen was an

exploratory, descriptive design. The purpose of exploratory studies has been described as being ' ... to gain familiarity with a phenomenon or to achieve new insights into it, often in order to formulate a more precise research problem or to develop hypotheses.' (Selitz 1965:50). Besides Leedy's concept of the nature of the data dictating the research method, there are various other aspects which affect the choice of research method and data collection strategies. A range of constraints and practical problems in the present study affected the methodology (described in the data collection section - see p 76). There are various problems associated with exploratory studies, eg. overload of information, but as Yates et al point out: ' ... there is no perfect study design, only an array of optimal choices.' (1983:2378)

The present study involves a survey of 38 adults who had been born with a congenital anomaly that required surgical correction.

5.2 SAMPLING

A sample is a group of subjects selected from a larger group including less than all the subjects in the larger group (Polansky 1975).

A. Demarcation of the study

The population of study was limited to all patients born prior to 1966 inclusive, admitted to The Children's Hospital, where a diagnosis of Hirschsprung's disease, oesophageal atresia or high anorectal malformation had been made. This included patients who had been diagnosed and operated on as emergency cases at other hospitals, but whose definitive corrective surgery was performed at The Children's Hospital.

It must be noted that while current hospital records are computerized, records from the period under discussion are probably inaccurate. As the hospital only opened in 1956, some of the respondents had been operated on initially at Groote Schuur Hospital and often had definitive surgery and post-operative treatment at The Children's Hospital. This could account for omissions and inaccuracies in hospital records. While the writer has attempted to obtain accurate records, the above limitations must be borne in mind.

The universe was 302 patients (44 OA patients, 107 HD patients, 151 AM patients). However, after patients who were not suitable for the study had been excluded (188), 114 patients were potentially available for the study.

B. Exclusions

- (a) As one of the aims of the study was to compare the effects of severity of disability, only high anorectal malformations were studied and low anomalies, eg. vestibular anus or covered anus were excluded as these are less severe forms of disability, and the anorectal malformation group was selected to represent the most severe congenital anomaly group in the study.
- (b) All patients presently living outside the Republic of South Africa were excluded, as the practical difficulties in tracing them and using them in the study were too great.
- (c) All 'African' patients were excluded because of language and tracing difficulties.
- (d) All those with other associated congenital anomalies were excluded as it was felt that the effects of the congenital anomaly under discussion would not be able to be isolated from

the effects of other anomalies.

- (e) All those who were mentally impaired were excluded, as problems were anticipated in obtaining the kind of information required in this study.
- (f) All those with permanent colostomies were excluded, as it was felt that such patients had a more visible disability than those without colostomies.
- (g) Obviously, all those who had died prior to this study were excluded. Table 1 shows the total number of exclusions and the total number of patients and Tables 2, 3, 4, show the breakdown of exclusions according to diagnosis. The latter breakdown is important as one can see that, for example, in the oesophageal atresia group half of the patients died.

TABLE 1 TOTAL NO OF PATIENTS AVAILABLE FOR STUDY (ALL GROUPS)

Total no of patients	302
Total no of exclusions	188
Total no of patients available for study	114

TABLE 2 NO OF PATIENTS AVAILABLE FOR STUDY (HIRSCHSPRUNG'S DISEASE)

Total no of patients	107
No of patients died	23
No of patients not in SA	9
No of patients with permanent colostomies	2
No of patients with other anomalies	2
No of mentally impaired patients	3
No of Xhosa-speaking patients	6
Total no of exclusions	45
Total no of patients available for study	62

TABLE 3 NO OF PATIENTS AVAILABLE FOR STUDY (HIGH ANORECTAL MALFORMATIONS)

Total no of patients	151
No of patients died	23
No of patients not in SA	3
No of patients with permanent colostomies	5
No of mentally impaired patients	1
No of Xhosa-speaking patients	7
No of patients with low anomalies	78
Total no of exclusions	116
Total no of patients available for study	35

TABLE 4 NO OF PATIENTS AVAILABLE FOR STUDY (OESOPHAGEAL ATRESIA)

Total no of patients	44
No of patients died	22
No of patients with other anomalies	4
No of Xhosa-speaking patients	1
Total no of exclusions	27
Total no of patients available for study	17

C. Sampling method

The sampling method employed in this study was non-probability sampling. Kitson et al (1982) note that this type of sampling is used when the universe represented by the respondents, and the potential biases introduced by the sampling method, cannot be determined. This is definitely applicable to the present study.

Furthermore they note that this type of sampling may be the most appropriate for examining sensitive, relatively low frequency events (Kitson et al 1982). In the present study the events, ie children with the particular anomalies under discussion, are low frequency events.

D. Tracing difficulties

After the exclusions had been finalized the writer began tracing all the remaining 114 patients. This proved to be a difficult task as the last address which appeared on the folder was frequently the address of the family when the child was a few months or years old. The subjects are presently adults over 20 years old and many of the families have moved. Another difficulty was that some female patients had married and had changed their surnames. Some of the patients had been included in De Wet's (1984) study or had kept in contact with the Department of Paediatric Surgery at the hospital, so in these cases a more recent address was available.

The following tracing attempts were made: Firstly, letters were sent to the last address that was available for each patient. If this yielded no response, a letter was sent to the occupants of the last address enquiring whether they knew the patient and requesting that they contact their neighbours in this regard. If this yielded no response, the writer used the telephone directory and telephoned people with the same surname as the

patients in an attempt to discover the patient or a relative of the patient. In some cases the writer followed-up patients based on information in the patient's folder or that the surgeons had given her, eg. if the patient had recently contacted the surgeon. If none of the above methods yielded any response or the correct address, letters were sent to the patient for a second time. If after a month there was still no response, letters were sent to the patient for a third time. The tracing attempts described above took approximately 6 months. All active tracing ceased after this time, but if patients replied after this, they were included in the study.

Tracing proved to be a very time-consuming tedious process which yielded few responses. During the course of the research a few patients changed addresses or left the country and were therefore lost to follow-up. Table 8 shows the tracing record. It must be noted that 'no contact' refers to patients whose names were discovered too late in the study to begin the long tracing process. The reason they were not included initially was either because of an incorrect diagnosis or the fact that the patients were born prior to 1956. Originally the study was to include patients born in the 1956-1966 (inclusive) period, but a few patients who contacted the hospital with ongoing problems and who were born prior to 1956 were included in the sample as it was so small. However, their inclusion dropped the actual percentage traced from 50% to 43% as it was decided not to attempt tracing other patients born prior to 1956, as the difficulties increase with the older patients, and this change occurred at a late stage in the study. At one stage the writer considered including younger patients, eg. 16-20 years in the study to increase the sample size, but it was decided that this age group was dealing with different life issues and would not be comparable.

TABLE 5 NO OF PATIENTS TRACED : HIRSCHSPRUNG'S DISEASE

No of patients available for study	62
Letters returned addresses unknown	20
No reply to letters	9
No contact	9
No of patients traced	29

TABLE 6 NO OF PATIENTS TRACED : ANORECTAL MALFORMATIONS

No of patients available for study	35
Letters returned address unknown	13
No reply to letters	5
No contact	3
No of patients traced	13

TABLE 7 NO OF PATIENTS TRACED : OESOPHAGEAL ATRESIA

No of patients available for study	17
Letters returned address unknown	4
No reply to letters	3
No contact	3
No of patients traced	7

TABLE 8 PERCENTAGE TRACED

DIAGNOSIS	ORIGINAL SAMPLE	EXPANDED SAMPLE
Hirschsprung's disease	54% (n=53)	54% (n=62)
Anorectal malformation	32% (n=31)	31% (n=34)
Oesophageal atresia	14% (n=14)	15% (n=17)
TOTAL	50% (n=98)	43% (n=114)

It can be seen from the above tables that the anorectal malformation group yielded the lowest percentage that were traced. A possible reason for the difficulties in tracing them is that these patients might have moved away from their original communities, where it is very likely that their problem would have been well known from their childhood and adolescent years (especially at school), to live in areas where they could try and hide their problem. De Wet (1984) points out that some authors note that tracing difficulties may reflect mobility due to crisis. Another possibility is that severe physical problems had been viewed as unchangeable and as they grew up the embarrassment associated with seeking medical help had prevented their further contact with The Children's Hospital or any other hospital.

Bearing these tracing difficulties in mind and the small sample traced, it was seen to be useful to examine the meaning of these difficulties for the study and see how representative the sample was and speculate on possible biases.

E. Representativeness of sample and sample bias

As there was such a low percentage (43%) of patients traced, it was decided to see if there was a difference between the traced and not-traced group which could possibly explain why certain patients could not be traced. A chi-squared test was run using the 4 variables available from both groups - age, sex, race and diagnosis. No significant difference was found between the traced and not-traced patients and it was decided that within the context of the many constraints that the sample was adequate for the study.

TABLE 9 CHI-SQUARED ANALYSIS : TRACED COMPARED TO NOT TRACED

VARIABLE	χ^2	DF	P
Sex	0,0498	1	0,05
Age	1,1902	2	0,05
Race	0,5523	1	0,05
Diagnosis	0,6908	2	0,05

Kitson et al (1982) point out that it is important to report on data comparing the characteristics of respondents and non-respondents as this information is needed for drawing conclusions concerning potential biases in the sample, especially when the sample coverage is low. They include untraceable patients in their description of non-respondents (refusals are discussed under data collection - see p 76).

It is difficult to determine in which way the sample could have been biased. As regards severity, those with very severe problems might have kept more contact with hospitals and clinics. On the other hand, it was found that those patients who were traced but did not respond or refused to participate in the study, did have severe physical and psycho-social problems. Therefore, those that did not reply or sent back letters 'address unknown' might have been in the more severe group. De Wet (1984) points out that in some of the severe cases family disintegration or total abandonment of the patient might have occurred, and these patients would have been more difficult to contact.

Referring to less severe cases, those with few problems might not have bothered to respond as they were having so few problems, or they might have responded because they did not feel threatened taking part in a research study. Therefore it is not clear whether severity was a bias in the sample.

Other sample biases emerged concerning the vast differences between individuals

in the sample. Watson and Kendall (1983) note that several authors point out that chronic-disease patient groups are often mistakenly viewed as single, homogeneous populations, whereas they are usually heterogeneous groups that include individuals who differ greatly with respect to a variety of demographic characteristics. Viewing the patients as a relatively homogeneous group by virtue of the fact that they were born with the same illness certainly proved to be a problem in this study. As the study progressed the sample was found to be very varied and this caused a range of data collection difficulties.

Watson and Kendall (1983) feel that a sample of patients seen at a single site may not constitute a representative sample of a total chronic disease population and have coined the term 'sample of convenience' to describe this sampling method. To a large extent the present study used convenience sampling. However, many patients were visiting different hospitals and doctors and although most of them were originally operated on at The Children's Hospital, they were not all drawn from a single site.

F. Conclusions concerning sampling

"A bad sample is a bad sample. It cannot be dressed up and treated as more than it really is. It can however be adequately described and used within the limits of its generalizability" (Kitson et al 1982:978).

It is concluded that the sample used in the present study has various limitations which have been described. The author must emphasize that generalizability to other patient groups born with this anomaly may be limited.

5.3 DATA COLLECTION

Orr et al (1984:152) note that 'no single tool for measuring psycho-social function exists'. There are numerous psychiatric rating scales, personality inventories (eg. CPI; MMPI), projective techniques (Rorschach Test, T.A.T.), scales assessing social adjustment (Weissman 1975) and scales assessing psycho-social adjustment to illness (Morrow et al 1978). However, no single measure has been found to be adequate for psycho-social measurement. Orr et al (1984) note that the solution for many researchers is to use multiple measures, each assessing different psychological concepts in an attempt to cover the broadest possible areas of functioning. Furthermore, as Watson and Kendall (1983) point out, the use of these tests (eg. MMPI) with chronic disease patients is questionable because they include many items reflecting persistently poor physical health and reduced mobility.

The present exploratory study attempted to identify the impact of illness/disability on patients' psycho-social functioning. As this is such a new area it was not known which aspect of a person's life would be most affected by illness and so an attempt to examine all major aspects of a patient's life was initially embarked on. For this reason an eclectic data collection strategy was used and a few different measures were used. Various problems that arose during the course of the study, eg. illiteracy, provided other reasons for data collection methods to be modified and other methods used, which was another important reason for using multiple measures.

The data collection methods used were a combination of descriptive survey and the case study method. The descriptive survey specifically used questionnaires and the case study used structured interviews.

A. Decisions concerning data collection

Initially it was decided that the main research method to be used would be the self-administered questionnaire. The reasons for this were that not all the subjects lived in Cape Town and the subject matter is very sensitive, embarrassing and emotion-laden. Referring to the latter the author is in the same age category as the subjects and is female, whereas most of the subjects are male. After discussions with the Human Sciences Research Council it was initially decided that the author's age and sex combined with the very sensitive subject matter, eg. bowel and sexual aspects, would not provide an interview situation conducive to obtaining valid information. Kitson et al (1982) note that not enough attention is given to the characteristics of those collecting the data in research studies. Although various researchers note that interviewing is a more appropriate way of revealing information that is sensitive and complex, it was decided that in this study self-administered questionnaires would allow subjects a freer environment to write about sensitive issues. DeLamater (1982) points out that there has been concern that anxiety-arousing topics elicit false responses and he notes that Sudman and Bradburn's (1974) review of the literature substantiates this by noting that threatening questions elicit a substantial effect on response. They note that one of the most threatening areas is that concerning major health problems. It was therefore decided that questionnaires would be used.

The literature identifies a range of advantages and disadvantages of questionnaires and in this study the self-administered questionnaire is specifically referred to. Advantages of questionnaires include being less expensive to administer as they can be mailed and there can

be anonymity if unsigned questionnaires are used. Mailing was used extensively in the present study. All questionnaires were coded by a research number and the respondent's name was not used on the actual questionnaire, although the author did contact some subjects personally on the telephone and visited some of them, which would have reduced anonymity.

Disadvantages include the problem of response rate. Polansky (1975) notes that a mailed questionnaire will not get a high response rate unless there is a strong incentive for the respondent to comply. In some cases in the present study respondents did reply because they needed help with social or physical problems. However, other patients who did not have these problems did not have strong incentives to reply. Furthermore perhaps those with extensive physical and/or psycho-social problems might not have replied either, probably as their incentive was lowered by the threatening nature of questionnaires. There was definitely a problem with response rate in the present study.

B. Problems with data collection

Another problem arose in the early stages of the study. Through pilot study contacts and letters to patients requesting them to fill in a simple form, eg. marital status, address, the author noted that many patients seemed to have literacy problems. Polansky (1960) notes that the ability to complete self-administered forms depends on the literacy of the respondent and with respondents of low education levels the use of these instruments is limited. A pilot questionnaire was therefore designed (Questionnaire A - see Appendix 2) to assess literacy and also to obtain some basic information - demographic and

family background information. The author tried to deliver as many of the questionnaires personally as possible in order to explain the purpose of the research study and to have an informal talk with patients to build up rapport. A second questionnaire (Questionnaire B - see Appendix 3) was administered at the same time. This was the Clinical Measurement Package which measures problems in various aspects of psycho-social functioning. Where the author suspected there might be a literacy problem she stayed with the patient while he/she filled in the questionnaire and helped clarify certain questions. When questionnaires were sent back and it was obvious the subject had not understood the questionnaire, the author phoned the subject and clarified answers telephonically. This does in some way bias the procedure as the writer had personal contact with some patients and not others. However, the extremely varied sample, ie. social class, educational levels, necessitated this compromise and in fact many other modifications in the study.

After Questionnaire A and B had been completed the writer had a fairly good idea of who could answer questionnaires adequately and in administering the final questionnaire (Questionnaire C - a medical questionnaire - see Appendix 4), similar methods were used as with the other questionnaire, ie. the writer was present or asked the questions herself with those who could not adequately answer questionnaires.

Towards completion of the study, it was decided that more in-depth information could supplement the existing information and so one case was chosen from each anomaly group for a case-study. Runyan (1982) points out that while the case study method has been criticized for a lack of controls, for inadequate measurement of independent and

dependent variables and for data being interpreted in an arbitrary manner, the richness of material cannot be obtained in any other way. He also notes that the case study is primarily useful for tasks such as describing an individual's experiences. As this was the type of information required in this study, the case study method seemed to be a viable research option. Although there were the difficulties concerning the writer's age and sex (already discussed), it was decided that the in-depth material needed in the case studies could only be obtained through interviews. It was also felt at this stage that the writer had gained some rapport through the initial phases of the research project, that she was seen as part of a medical team and that in answering the medical questions in Questionnaire A perhaps some of the initial defences regarding embarrassing questions had been broken down. This research method, ie. interviewing, created freedom for respondents to express thoughts and feelings, but as with all interviews, interviewer bias must be taken into account during the interview and in the interpretation of data.

Another problem concerning data collection was that of refusals. No respondent directly refused to answer the questionnaires, but one respondent's mother phoned to say that he had received the questionnaire, but whenever she asked him if he had sent it back, he became angry and he in fact did not send it (he had Hirschsprung's disease). One patient's employer, a qualified nurse, had tried to encourage another patient to answer the questionnaire but he refused (he had an anorectal malformation). Both these patients were seen to have physical problems according to those closest to them, so possibly the questionnaire was too threatening. Other respondents kept promising to send the questionnaire back, but did not in fact do so

and it is not clear whether this was for the reason proposed above or for another reason.

C. Methods of analysis

All the measures used were self-report measures which although problematic in some ways, eg. people under or over-reporting behaviour, feelings, they do, according to some authors (Watson and Kendall 1983), retain an important place in research as there are no good alternatives for assessing certain types of constructs. These assessment techniques measure respondents' perceptions regarding behaviour and feelings.

Three questionnaires were administered to all patients (descriptive survey method) and three patients (one from each anomaly group) were interviewed (case-study method).

(a) Questionnaire A (see Appendix 2)

The aim of this questionnaire was to test literacy and obtain demographic and family background information. Questions included closed (forced-choice) and open-ended questions. There was a section for any other comments at the end that allowed free expression of thoughts and feelings. Many of the subjects whom the writer met verbalised a great deal when asked for any other comments.

(b) Questionnaire B (see Appendix 3)

This questionnaire is called the Clinical Measurement Package and was designed by Hudson and published in 1982. It is an American scale which consists of 9 psycho-social scales and is aimed at identifying the extent of problems in various psychological dimensions, eg. self-esteem, general contentment (which measures depression) and

various social dimensions, eg. peer relationships, marital relationships. It consists of various statements and the respondent is given a choice of one out of five responses with which to answer each statement. Six of these scales were chosen for the study. The scale concerning a respondent's attitude towards their child was not included as only 9 of the respondents had children. The scale concerning the respondent's attitude towards his/her mother and his/her father was not included and was represented instead by the scale concerning family relationships. A high score on a scale indicates that the respondent has a problem with that aspect of his/her life. The writer added in an explanation on some scales concerning which scales were applicable to respondents to clarify which ones they should answer, eg. the sexual satisfaction scale was only to be answered by respondents who had had or were currently having a sexual relationship.

(c) Questionnaire C (see Appendix 4)

This questionnaire is a medical questionnaire aimed at assessing the subject's past and present health generally and specifically, referring to his/her problems relating to the anomaly. Residual problems were examined in the past and at present, as an effect on present psycho-social functioning could be caused by past residual problems, present residual problems, or both. All respondents were asked whether they were experiencing certain symptoms common to all groups in order to compare the groups adequately, eg. all subjects were asked the same questions. This was used as a type of control. For example, it was useful to see how many patients with oesophageal atresia suffered from constipation. This provided a norm with which to compare subjects with Hirschsprung's disease and anorectal

malformations. A funnel approach was used, ie. general to specific in that questions were initially asked in a general way to allow more freedom of response for the subjects to see how open they were and then more specific questions were asked to try to get information from those who did not respond to the open-ended questions.

(d) Questionnaire D (see Appendix 5)

Three subjects were selected for the case study and interviewed using an interview schedule consisting of semi-structured questions. This interview examined various aspects of the respondents' life in the past and in the present, eg. work, peers, school aspects. As with the medical questionnaire it initially gave the subjects space to give their impressions of how the illness affected their lives and then asked more specific questions in case there was little response initially. Overall the approach followed here was similar to what Tripodi and Epstein (1980) describe as the 'umbrella approach' in that instead of moving from general to the specific, the interviewer comprehensively covers all aspects of a given dimension.

DeLamater (1982) notes that questions about past events should be avoided whenever possible. It was felt in this study that past events would affect present psycho-social functioning so questionnaires elicited past and present information although it was found that patients frequently did not remember past events.

The questionnaires used in the study were piloted on various members of the public (medical and non-medical), and although the writer attempted to find a range of education levels those piloted tended to have achieved a fairly high education level which did not represent

the full range of education levels in the study sample. Nevertheless this piloting method did prove to be very useful in making various questions less ambiguous and achieving better clarity in general.

D. Procedure

Questionnaires A and B were either mailed with self-addressed envelopes requiring no postage, or dropped off personally by the writer. Attention was paid to ensure confidentiality in a covering letter and only numbers on questionnaires were used, not names. The covering letter included the aim of the questionnaire and the importance of research for future medical care in order to motivate subjects to respond. Goldstein (1969) notes that the worth of the research being done should be explained to the respondent.

Based on the responses to these questionnaires, Questionnaire C was designed and either sent to respondents or the writer went through the questionnaire with the respondent. It was found that many respondents initially reported no physical problems when the questions were asked in a general and open-ended manner, but later they admitted to problems when more specific questions were asked. Questionnaire C identified many respondents who were having residual symptoms and the writer was involved in clinical work with some respondents and referred them for psychiatric and medical assessment and treatment.

Two follow-up letters and 2 telephone reminders were used to encourage respondents to reply. If after this the respondent had not replied he/she was excluded from the study. There were problems with questionnaire response rates as mentioned earlier. Kitson et al (1982) note that unless vigorous follow-up techniques are used with mailed questionnaires, which was done in the present study, response rates are usually low. Table 10

shows the number of patients who responded to the questionnaires and were included in the study, ie. the final sample.

TABLE 10 RESPONDENTS INCLUDED IN THE STUDY (FINAL SAMPLE)

	N	%
Oesophageal atresia	5	13
Hirschsprung's disease	24	63
Anorectal malformations (high)	9	24
TOTAL	38	100

Three respondents were selected from each anomaly group for the case studies. It was decided to select a respondent who had many residual problems in each group so as to obtain a clearer indication of the range of problems afflicting respondents and the possible psycho-social effects these problems were having. Selection of respondents was based on data from the medical questionnaire (Questionnaire C).

The case study involved a 2 - 3 hour interview. During the interview, probing was used to elicit responses or to clarify questions. After the interview the writer wrote down the information immediately afterwards as it was felt it would be too distracting within the interview situation. Tripodi and Epstein (1980) note that sometimes the interview situation is so highly charged, it would be impossible or grossly insensitive for the interviewer to be writing while the respondent is revealing his/her most intimate problems, thoughts and experiences.

5.4 DATA ANALYSIS

A. Primary analysis: (of data from Questionnaire A,B and C)

(a) Analysis of Questionnaire A

Information was divided up according to the questions and sections in the questionnaire. It was further subdivided according to diagnosis. Information was recorded in frequencies and not percentages as it was decided that the latter were meaningless in view of the small sub-samples. As the sub-samples were small and to facilitate analysis, categories were collapsed so that there were fewer subdivisions for each variable.

Further analysis was conducted on some variables to obtain an overcrowding index, a social problem index and a social class index. The questions concerning number of rooms in the house and number of people, were used to calculate the Batson index (Batson 1944) which gives an index of overcrowding in a house (see Appendix 6). The questions concerning parents' criminal offences, substance abuse, death and/or illness, divorce and marital difficulties, psychiatric problems and family violence, living arrangements and geographic mobility were used to decide whether a family had social problems or not. Living arrangements and geographic mobility were discarded as it was decided that these could not necessarily be seen as major determinants of social problems. If a respondent's family had two or more of the other social problems, or mentioned any other social problems, he/she was placed in the social problem group. All other respondents were put in the no social problem group.

For the purposes of the present study occupation of the

respondent and the respondent's family were analyzed according to the British Registrar General's Outline used for census purposes described in Davie, Butler and Golstein (1972:2-6) (see Appendix 7).

This classification system highlights five occupational groups which are described as social classes and range from the highest social class (social class I) to the lowest social class (social class V). All respondents were classified in this way. (If they were unemployed or housewives, their last occupation was taken; university students were put in social class I). As the father was the main breadwinner in almost every family, it was decided that the father's occupation (therefore his social class), would be used as a measure of social class of the respondent's family.

(b) Analysis of Questionnaire B

These scales were each scored according to the CMP scoring system. A value is obtained (for each scale) between 0 and 100 and this indicates the magnitude of a problem in this dimension. The cut-off point is 30 and all those with values greater than 30 are seen as having problems and all those with values less than 30 are not seen as having problems. Hudson (1982) suggest a cutting range of 25 to 35 where it is uncertain whether a person has problems or not. All the respondents were divided on each scale according to a problem group, an uncertain group and a no problem group. This information was then examined according to diagnosis and severity (past and present). The information was recorded in frequencies.

(c) Analysis of Questionnaire C

All the information was divided up according to the questions and sections in the questionnaire, recorded in frequencies and subdivided according to diagnosis. Some variables were also divided according to severity.

B. Secondary analysis (of data from Questionnaire A,B and C)

Data was analyzed descriptively.

The writer examined the available non-parametric statistical tests which would examine the difference between independent samples using nominal data. The Chi-squared test was the appropriate test, but had to be rejected as the sample size was too small. Unfortunately the Yates' test (a correction of the Chi-squared test) and the Fisher Exact Probability test which would have accommodated the small sample size, could only be used for data in 2x2 tables. As this study involved comparing data between 3 independent groups, these tests could not be used.

Therefore, as there was no appropriate statistical test, and bearing in mind that small samples do not lend themselves to statistical analysis as well as large samples do, it was decided to analyze all the data descriptively.

Each variable was displayed graphically according to diagnosis and important variables according to severity (past and present) as well. It was decided to use this method as the data was in a frequency, not percentage format, and to compare groups in a graphical format was seen to be more useful.

When the data could not be displayed graphically, written descriptions

were given.

C. Analysis of Questionnaire D

The case studies were descriptively analyzed according to the subsections in the interview schedule and presented according to these subsections.

5.5. CONTROL GROUP

Initially the study of the oesophageal atresia anomaly group had a dual purpose. It was included in the study to represent a congenital anomaly group with few or no problems (mild severity) and it was included as a control group. It was selected to represent a group of patients who had experienced the early traumas of surgery, hospitalization and separation from parents, but after this had led a relatively normal life. If they had experienced residual problems these were likely to be mild and less socially handicapping than the residual problems associated with Hirschsprung's disease and high anorectal malformations. However, during the study it was decided that the sample for this group was too small ($n=5$) for this group to be used as a control group and therefore the study did not have a control group. Burish and Bradley (1983) note that lack of controls is a common problem in research concerning chronic disease.

5.6 PILOT STUDY

As the sample was so small the author felt she could not use any of the patients for a pilot study that were in the main study. To overcome this problem, one patient from each anomaly group was informally interviewed to gain a preliminary understanding of the sort of issues that were important. Furthermore, the writer attempted to pilot her research instruments.

(see p 83)

5.7 RELIABILITY AND VALIDITY

A. Reliability

Questionnaire data (for Questionnaire A, C and D) did not lend itself to the usual reliability checks, eg. test-retest method, split-half method, as these checks require questionnaires to contain numerical data and questionnaire data was often not in numerical form and sometimes in the form of open-ended questions.

The fact that the same researcher designed and administered all questionnaires and interviews does provide a certain amount of standardization of research items and consistency of methods.

The Clinical Measurement Package (Questionnaire B) has been checked for reliability by Hudson (1982), and he reports that these checks provide strong and convincing evidence that each of the CMP scales is a highly reliable measurement device. Reliability data is not yet available for one of the scales, the IPR scale (peer relationships). He notes that it is strongly suspected that it will be found to be a highly reliable device, taking into account the results of reliability checks for the other scales, but he suggests that results of this scale be treated with caution until reliability findings are made available.

B. Validity

The questionnaire data (for Questionnaires A, C and D) can be seen to have content and face validity. Based on the pilot study, De Wet's (1984) study and the literature, the writer designed the research instruments. It seems that these instruments are measuring what they intended to measure. However, as Nelson (1980) points out, content

validation lies basically in the judgment of the researcher, so is a subjective interpretation of validity. The questionnaires do not meet the requirements for other forms of validity.

Hudson (1982) reports that his validity checks provide reasonable evidence to support the claim that the CMP scales actually measure what they were designed to measure and can therefore be regarded as valid measurement instruments.

5.8 ETHICAL CONSIDERATIONS

The research study was accepted by the Ethics and Research Committee, Medical School, University of Cape Town. Permission was obtained from the publishers of the CMP to use this instrument in the research study.

As already discussed, attention was paid to ensuring the anonymity of respondents in the study by not using their names. Furthermore, the purpose of the research study and of the research questionnaires used in the study was explained to respondents at the various stages of the study.

5.9 LIMITATIONS OF THE STUDY

Limitations of the study included the small sample size, problems with sampling methods, ie. the sample not being randomly selected, some problems with the reliability and validity of research instruments, the lack of a control group and proper pilot study. However, taking into account the nature of the research and the practical and methodological difficulties, this study was seen to be valuable as an exploratory study, but with limited generalizability to other disease populations.

CHAPTER 6FINDINGS

This chapter presents the following information:

- 6.1 Diagnostic data
- 6.2 Medical and health data (see Appendix 9 for additional medical data)
- 6.3 Medical information and counselling data
- 6.4 Demographic data (see Appendix 8 for additional demographic data)
- 6.5 Socio-economic data concerning respondent (see Appendix 8 for additional socio-economic data)
- 6.6 Socio-economic data concerning respondents' families (see Appendix 8 for additional socio-economic and family background data)
- 6.7 Family problem data (see Appendix 8 for additional family problem data)
- 6.8 Psycho-social problem data (referring to respondents' current problems)
- 6.9 Information concerning the effect of the anomaly and residual problems on the respondent
- 6.10 Case studies - description of information received during interviews

6.1 Diagnostic data

Diagram 1 CONGENITAL ANOMALIES
Breakdown of Diagnostic Groups

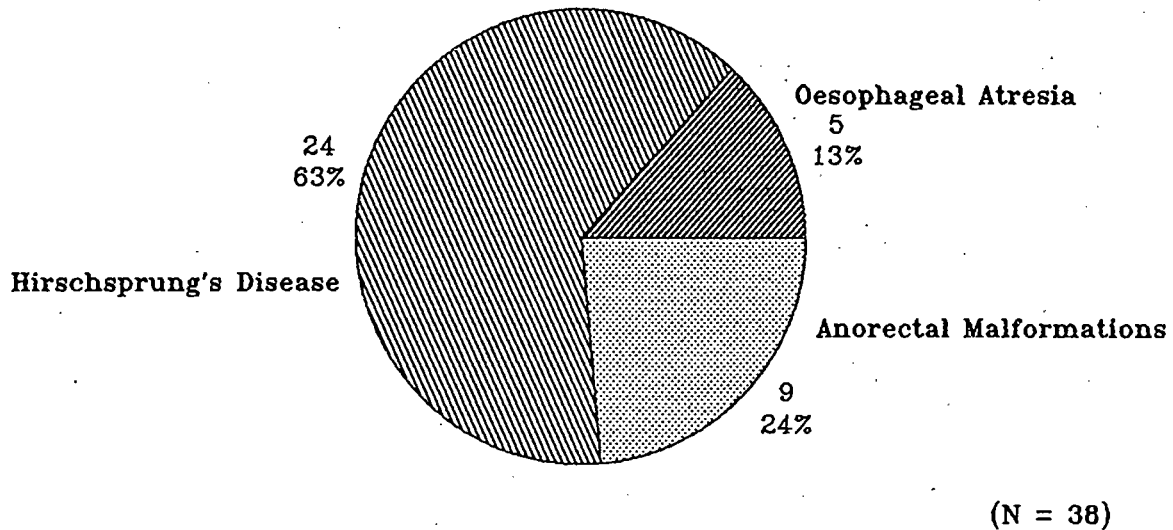
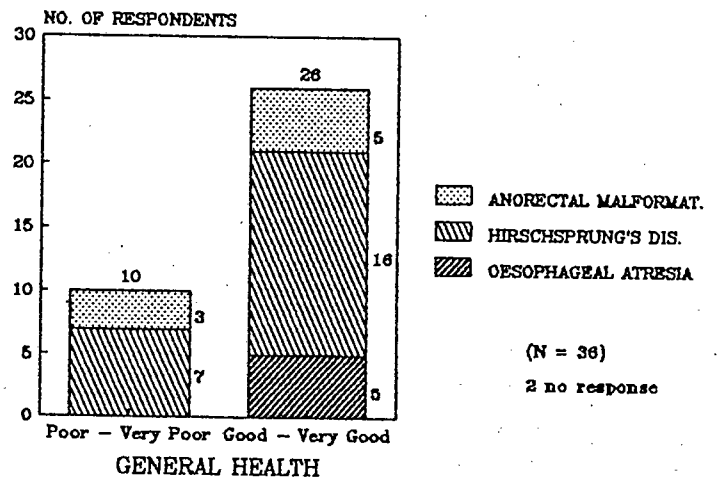
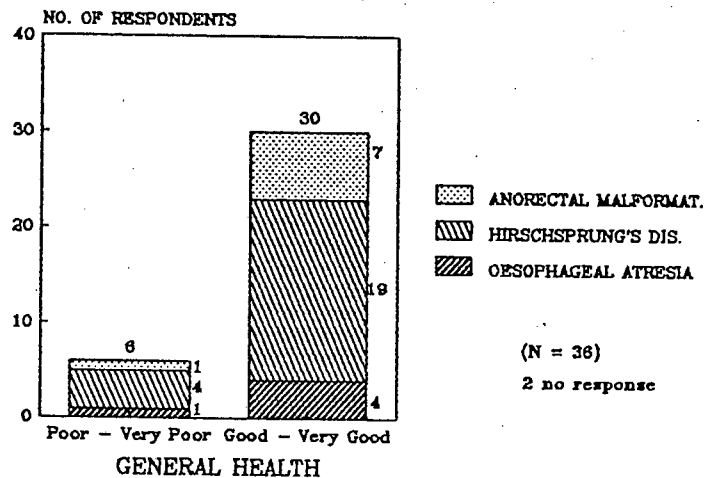


Diagram 1 indicates that HD respondents (24 out of 38) account for the largest proportion of the sample, AM respondents account for approximately one-quarter of the sample (9 out of 38) and OA respondents (5 out of 38) account for the smallest proportion of the sample.

(This graphical breakdown is placed at the beginning of this Findings section to clarify sample size, before other variables relating to anomaly groups are examined. It will not be discussed further in the Discussion section - Chapter 7).

6.2 Medical and health dataDiagram 2 DIAGNOSIS BY GENERAL HEALTH
(PAST)Diagram 3 DIAGNOSIS BY GENERAL HEALTH
(PRESENT)A. General health

To simplify interpretation of data, health was classified in terms of poor - very poor and good - very good. Diagram 2 shows that OA respondents (5 out of 5), HD respondents (16 out of 23) and AM respondents (5 out of 8) rate their health in the past as good to very good, although 7 HD and 5 AM respondents rated their health as poor to very poor. Diagram 3 shows that most OA respondents (4 out of 5), HD respondents (19 out of 23) and AM respondents (7 out of 8) rate their health at present as good to very good. Therefore HD and AM respondents tended to rate their health as better at present compared to the past. Although most OA respondents rated their present and past health as good to very good, 1 respondent rated her present health as poorer than in the past.

B. Nutritional and growth data

TABLE 11 MEAN ADULT STATURE (ALL GROUPS)

	Males	Females
Oesophageal atresia respondents	172	163
Hirschsprung's disease respondents	164	163
Anorectal malformation respondents	168	173

Normal mean height (males): mean = 174.3 cm

Normal mean height (females): mean = 163.8 cm

(a) Adult stature

The above table shows that the mean adult stature of male HD respondents was 164 cm and 168 cm for male AM respondents compared to a mean normal height of 174.3 cm (SD = 5,9), according to NCHS standards (1978).

The OA group and HD and AM female respondents had normal adult stature according to these standards. Adult stature is the outcome of statural growth until the closure of the epiphyses in late adolescence. Short stature of males with HD suggests malnutrition or some other factor interfering with childhood growth (only 1 HD male out of 16 had normal height). Short stature of males with AM might suggest malnutrition in the past, but as the sample size for AM respondents is so small mean measures are not as significant as for HD respondents.

TABLE 12 MEAN ADULT NUTRITIONAL STATUS (ALL GROUPS)

	Males	Females
Oesophageal atresia respondents	24	22
Hirschsprung's disease respondents	25	22
Anorectal malformation respondents	22	21

Normal mean Quetelet index (males) = 20,08

Normal mean Quetelet index (females) = 21,05

(b) Adult nutritional status

The above table shows that according to Quetelet indices (Woods 1984) all groups appear adequately nourished at present (Quetelet index = w/h^2).

Therefore all groups had normal height and weight, except for HD males and some AM males who had short stature.

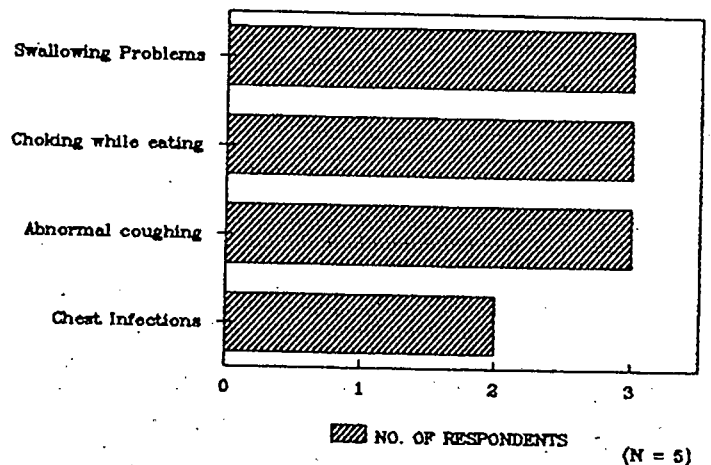
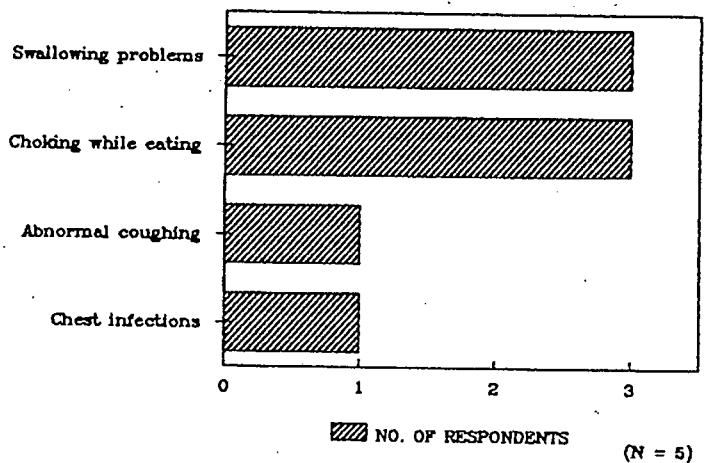
C. Residual problemsDiagram 4 RESIDUAL PROBLEMS (PAST)
Oesophageal AtresiaDiagram 5 RESIDUAL PROBLEMS (PRESENT)
Oesophageal Atresia(a) Residual problems (OA)

Diagram 4 reveals that the residual problems experienced by OA respondents, in the past, in descending order of frequency (according to the number of respondents who experienced a certain problem) include swallowing problems (3 out of 5), choking problems (3 out of 5), abnormal coughing (3 out of 5) and chest infections (2 out of 5). Diagram 5 indicates that the residual problems experienced by OA respondents at present include swallowing problems (3 out of 5), choking while eating (3 out of 5), abnormal coughing (1 out of 5) and chest infections (1 out of 5). Therefore OA respondents experienced fewer residual problems at present than in the past.

Diagram 6 RESIDUAL PROBLEMS (PAST)
Hirschsprung's Disease

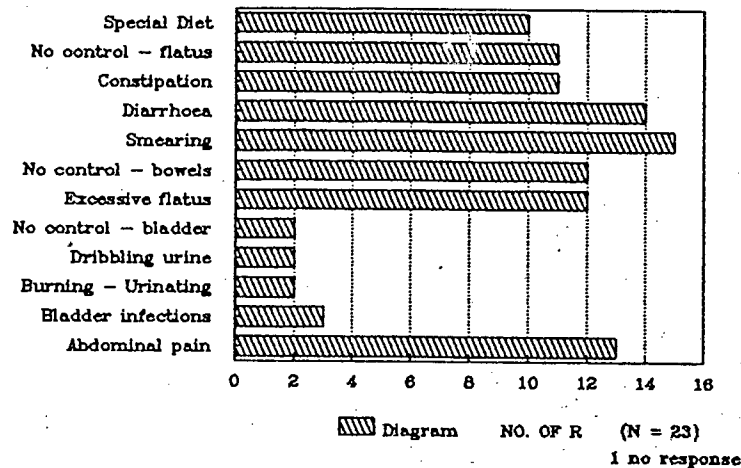
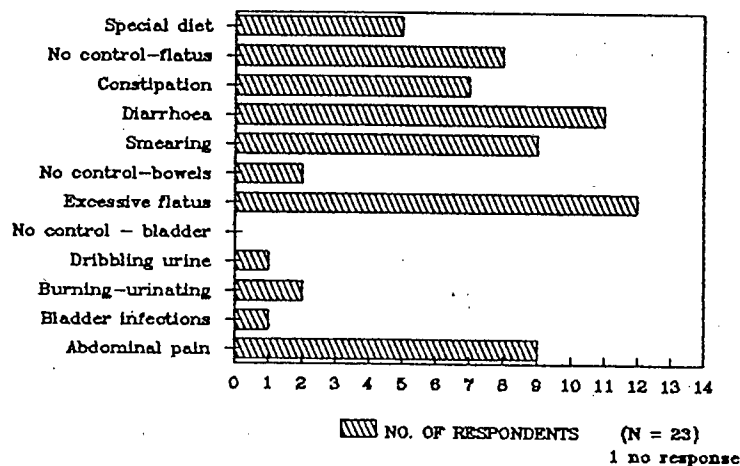


Diagram 7 RESIDUAL PROBLEMS (PRESENT)
Hirschsprung's Disease



(b) Residual problems (HD)

Diagram 6 indicates that the residual problems experienced by HD respondents, in the past, in descending order of frequency (according to the number of respondents who experienced a certain problem) include smearing (15 out of 23), diarrhoea (14 out of 23), abdominal pain (13 out of 23), no control over bowels (12 out of 23), excessive flatus (12 out of 23), no control over flatus (11 out of 23), constipation (11 out of 23), special diet (10 out of 23), bladder infections (3 out of 23), no control over bladder (2 out of 23), dribbling urine (2 out of 23), and burning when urinating (2 out of 23).

Diagram 7 (see p 97) indicates that the residual problems experienced by HD respondents, at present, include excessive flatus (12 out of 23), diarrhoea (11 out of 23), smearing (9 out of 23), abdominal pain (9 out of 23), no control over flatus (8 out of 23), constipation (7 out of 23), special diet (5 out of 23), burning when urinating (2 out of 23), no control over bowels (2 out of 23), dribbling urine (1 out of 23) and bladder infections (1 out of 23). Therefore HD respondents experienced fewer residual problems (bowel and urinary problems) at present. (Some respondents experienced more residual problems at present, as they have sexual and/or fertility problems - see p 101 and 102).

Diagram 8 RESIDUAL PROBLEMS (PAST)
Anorectal Malformations

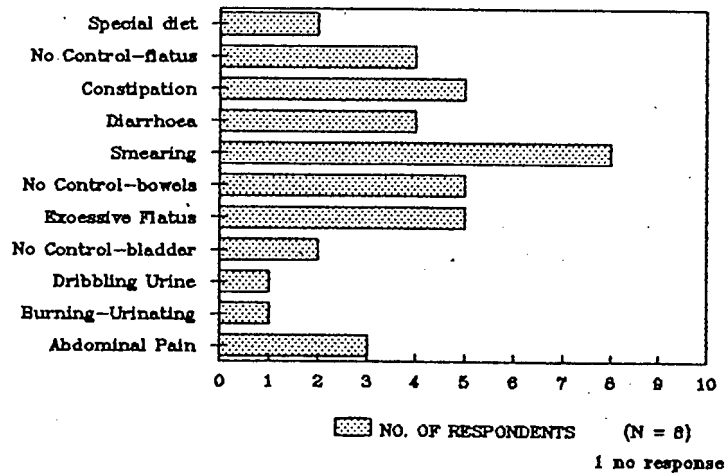
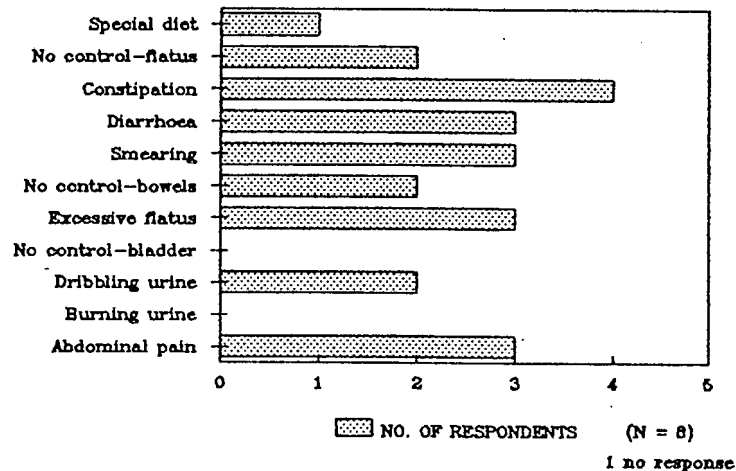


Diagram 9 RESIDUAL PROBLEMS (PRESENT)
Anorectal Malformations



(c) Residual problems (AM)

Diagram 8 indicates that the residual problems experienced by AM respondents, in the past, in descending order of frequency (according to the number of respondents who experienced a certain problem) include smearing (8 out of 8), constipation (5 out of 8), no control over bowels (5 out of 8), excessive flatus (5 out of 8), no control over flatus (4 out of 8), diarrhoea (4 out of 8), abdominal pain (3 out of 8), special diet (2 out of 8), no control over bladder (2 out of 8), burning when urinating (1 out of 8) and dribbling urine (1 out of 8).

Diagram 9 indicates that the residual problems experienced by AM respondents, at present, include constipation (4 out of 8), abdominal pain (3 out of 8), excessive flatus (3 out of 8), smearing (3 out of 8), diarrhoea (3 out of 8), no control over flatus (2 out of 8), no control over bowels (2 out of 8), dribbling urine (2 out of 8) and special diet (1 out of 8). Therefore AM respondents experience fewer residual problems (bowel and urinary problems) at present. (Some respondents experience more residual problems at present, as they have sexual and/or fertility problems - see p 101 and 102)

(d) Comparison of diagnostic groups' residual problems

OA respondents tend to have mild residual problems, in the past and at present, compared to the HD and AM respondents. In the past, AM groups had proportionately more bowel problems than HD respondents. The latter groups have a similar proportion of respondents with urinary problems. At present, the AM group tends to have a similar number of respondents (proportionately) with bowel problems, although more AM respondents to the HD group (proportionately) have severe bowel problems, ie no control over their bowels. (The HD and AM groups also have a similar proportion of respondents with residual problems relating to sexuality and fertility - see p 101 and p 102). Therefore OA respondents had, and still have, the fewest and mildest residual problems. The HD and AM groups, had and still have, a similar number of respondents (proportionately) with residual problems in the past and present, although the AM respondents, had and still have, more severe problems than HD respondents. An insignificant number of respondents reported residual problems common to other groups.

Diagram 10 DIAGNOSIS BY SEXUAL PROBLEMS

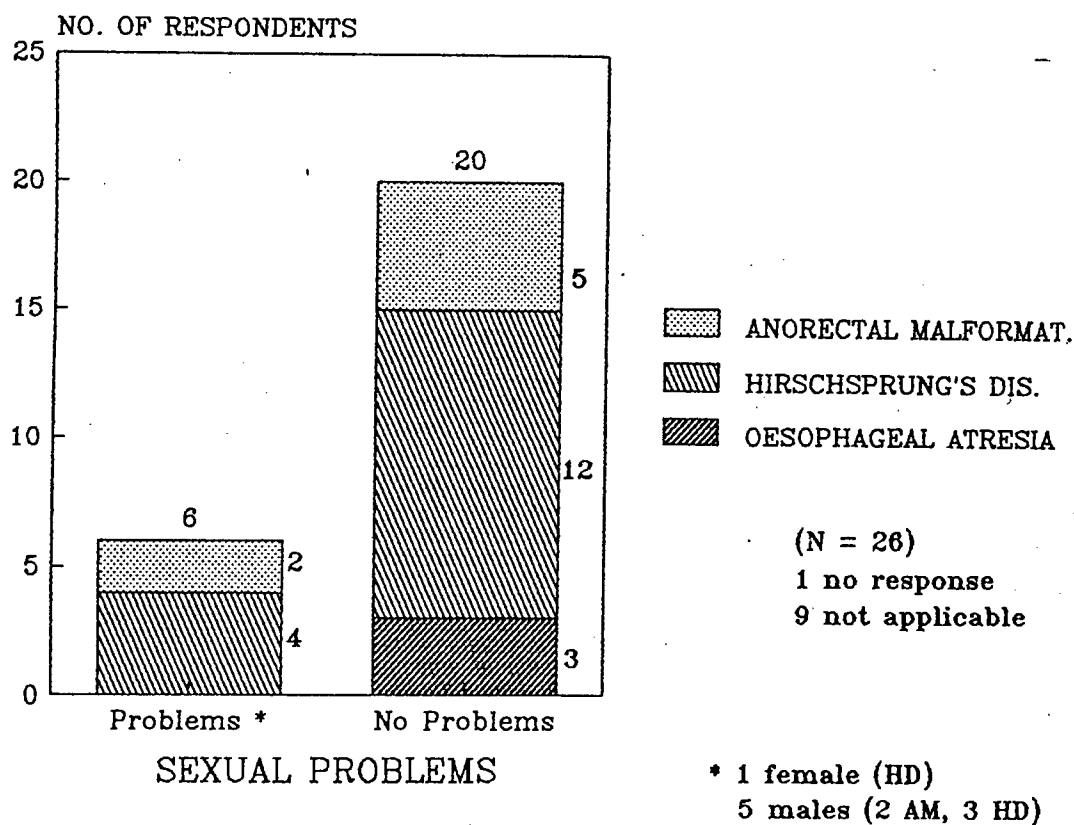
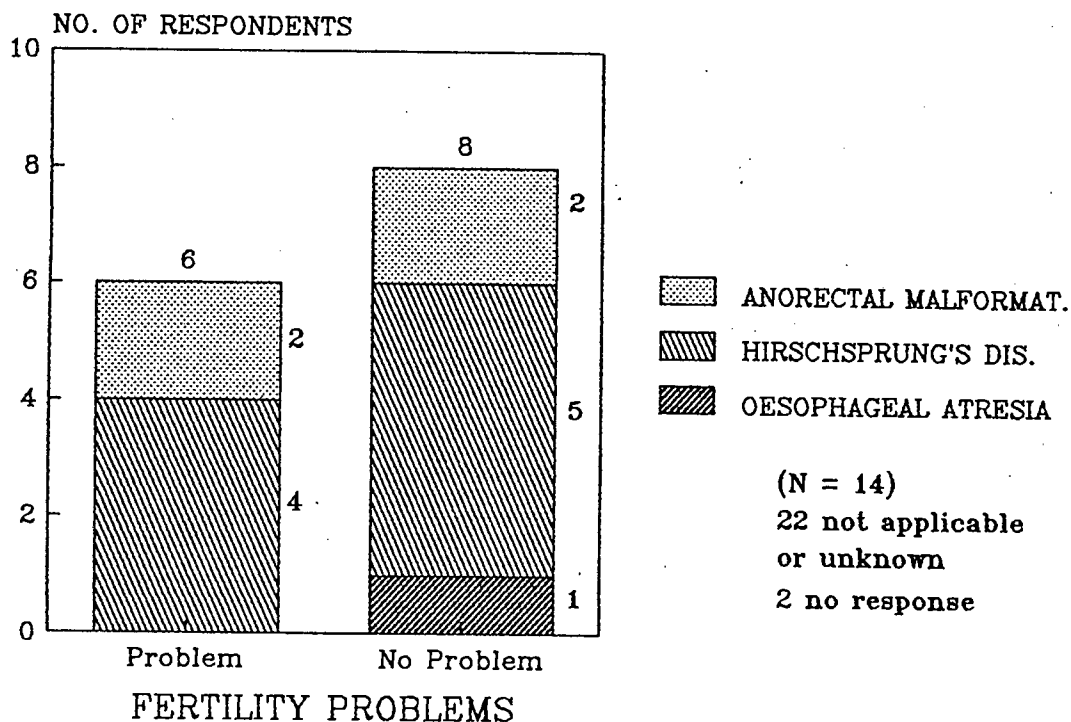
(e) Sexual problems

Diagram 10 suggests that there is a tendency for OA respondents (3 out of 3), HD respondents (12 out of 16) and AM respondents (5 out of 7) to have no sexual problems, although 4 HD respondents and 2 AM respondents have problems. (Through contact with one of the 2 respondents who did not respond to questionnaire C, it was discovered that this male HD respondent was having sexual problems and was included in the description below)

Type of sexual problems - Mucous discharge from anus during sex (1 male AM), pain during sex (1 female AM), difficulty with ejaculation (1 HD) and no ejaculation (3 HD, 1 AM). Therefore the HD and AM groups have a few respondents with sexual problems (similar proportion in the 2 groups), while OA respondents experienced no sexual problems.

Diagram 11 DIAGNOSIS BY FERTILITY PROBLEMS



(f) Fertility problems

Diagram 11 shows that the OA respondent (1 out of 1) did not have a problem with fertility. Although more HD respondents did not have a problem with fertility (5 out of 9), 4 did have problems and some (2 out of 4) AM respondents had fertility problems. (Through contact with one of the 2 respondents who did not respond to questionnaire C, it was discovered that this male HD respondent was having problems with fertility and he was included in the description below)

Type of fertility problem - No ejaculation (3 HD, 1 AM), internal scarring prevents pregnancy (2 female HD, 1 female AM). Therefore the HD and AM group have a few respondents with sexual problems (similar proportion in the 2 groups), while OA respondents have no sexual problems.

D. Restrictions on respondents' lives (past)

Diagram 12 DIAGNOSIS BY RESTRICTIONS
ON LIFE (PAST) *

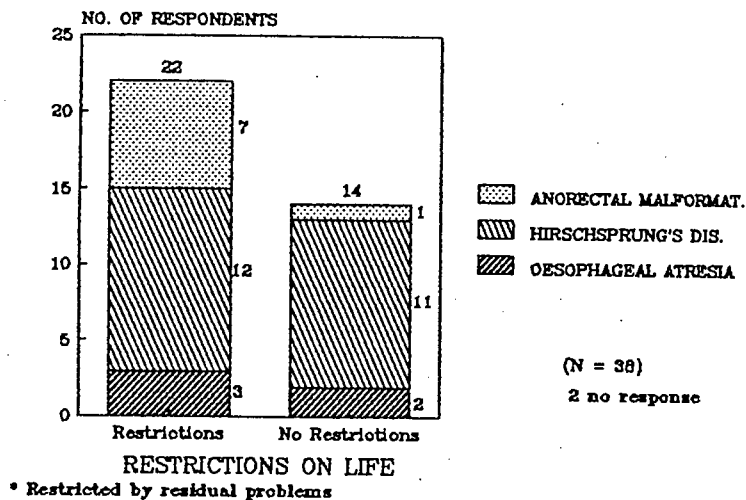


Diagram 12 shows that OA respondents (3 out of 5), HD respondents (12 out of 23) and AM respondents (7 out of 8) report that their residual problems restricted their lives in the past, especially the AM respondents.

Ways in which residual problems restricted respondents lives (in the past)

Could not go away on holiday (3 HD); could not sleep overnight at friends (4 HD, 2 AM); could not swim (1 HD, 2 AM); sport restricted (1 OA, 8 HD, 2 AM); social embarrassment over teeth discoloured from medication (1 OA); could not go out with friends to all the places they went (2 AM); found it difficult to mix with people (1 HD, 1 AM); could not eat everything (1 AM); swallowing difficulties meant forcing food down or leaving the table to vomit (1 OA); having little energy restricted social events (1 OA, 2 HD, 2 AM); problems with being teased at school because of the smell or marks on underpants (1 HD, 1 AM); embarrassment over scars when on the beach (1 HD); wearing a one-piece bathing costume (1 HD) and for another wearing a jersey on the beach (1 HD).

E. Restrictions on respondents' lives (present)

Diagram 13 RESTRICTIONS ON LIFE
(PRESENT)

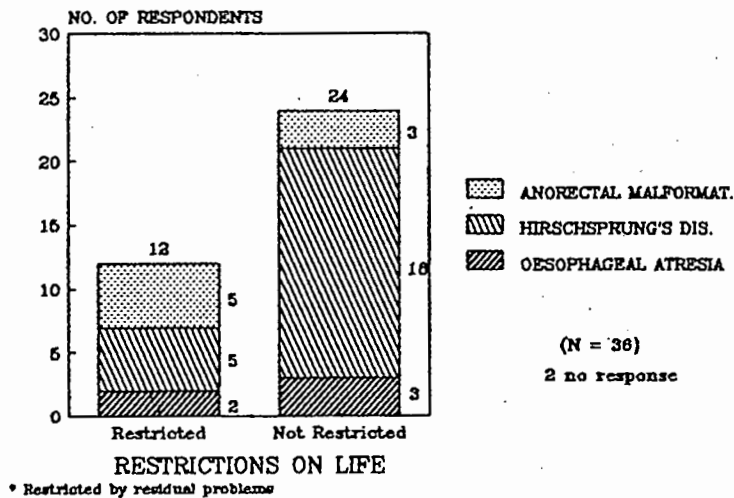


Diagram 13 indicates that the OA respondents (3 out of 5) and HD respondents (18 out of 23) report that their residual problems do not restrict their lives at present, whereas more AM respondents (5 out of 8) report that their residual problems presently restrict their lives.

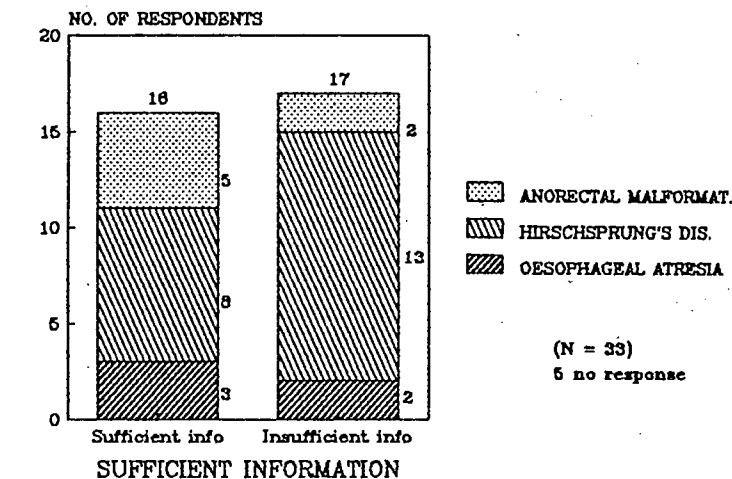
Ways in which residual problems restricted respondents' lives (at present)

Cannot play sport (2 HD, 2 AM); cannot mix easily (2 HD, 1 AM); cannot get a girlfriend (1 HD, 1 AM); restricts social activities (3 HD, 1 AM); embarrassing smell in public, especially when the weather is hot (1 AM); absences from work (1 HD, 1 AM); affects choice of occupation, ie must be non-manual (1 HD, 2 AM); cannot drink alcohol (1 AM); could not go to the army (1 HD).

Therefore all groups reported that their residual problems restricted their lives in the past, especially the AM group. At present, more OA and HD respondents report that their residual problems do not restrict their lives, whereas more AM respondents report that their residual problems do restrict their lives.

6.3 Medical information and counselling data

Diagram 14 DIAGNOSIS BY SUFFICIENT INFORMATION *



A. Medical information

Diagram 14 indicates that OA respondents (3 out of 5) and AM respondents (5 out of 7) reported receiving sufficient medical information concerning their anomaly whereas HD respondents (13 out of 21) did not feel they had received sufficient information.

Information needed by respondents (in their opinion) - Information was needed concerning the causes of the anomaly (4 HD, 1 AM); pathology (4 HD, 1 AM); explanation of operation (6 HD); how to cope with the residual problems (1 OA, 1 HD, 1 AM); long-term effects that the congenital anomaly would have (3 HD); possible treatments available (1 OA, 1 AM); which part of the body was operated on (1 HD); illustrations to show pathology and operation (2 HD); sexual/fertility problems (3 HD).

An additional comment by a respondent concerning information
 "... Enige ander persone wat dieselfde probleem het en nog baie jonk is, moet baie beter inligting gegee word oor al die nagevolge wat sy/haar lewe sal beïnvloed" (HD).

Age at which information was needed by respondents

How to cope with residual problems (adolescence - mentioned by 2 respondents); long-term effects (pre-puberty, adolescence, adulthood); causes (childhood, adolescence); illustrations (childhood, early adolescence); other treatments (adolescence); sexuality/fertility problems (pre-puberty, adolescence, late adolescence).

Respondents' answers for the part of the body which they think was operated on

Oesophageal atresia respondents (n=5)

Oesophagus (1); heart (1); no answer (1); tubes from lungs (2)

Hirschsprung's disease respondents (n=23 - 1 no response)

Stomach (14); colon (9)

Anorectal malformation respondents (n=8 - 1 no response)

Stomach (5); anus (3).

B. Hereditary aspects

Diagram 15 DIAGNOSIS BY RELATIVES WITH SAME ANOMALY

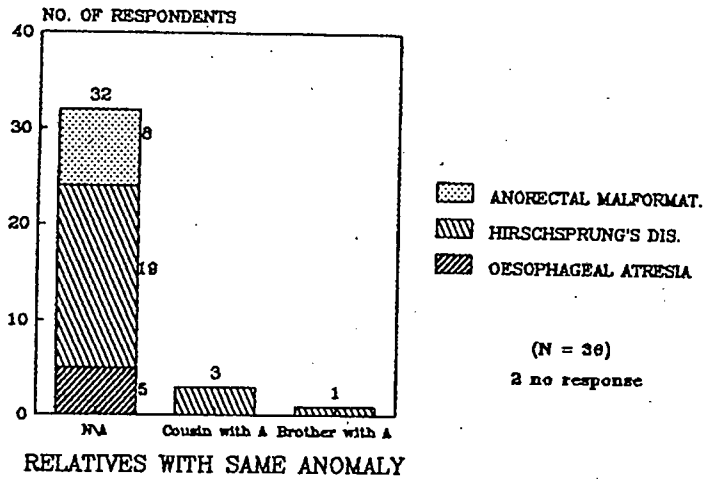


Diagram 15 indicates that no OA respondents (5 out of 5) and AM respondents (8 out of 8) had relatives with the same anomaly (as the respondent). Most HD respondents (19 out of 23) did not have a relative with the same anomaly, although 4 did - 3 had cousins and 1 had a brother with the same anomaly.

C. Counselling

Diagram 16 DIAGNOSIS BY NEED FOR COUNSELLING

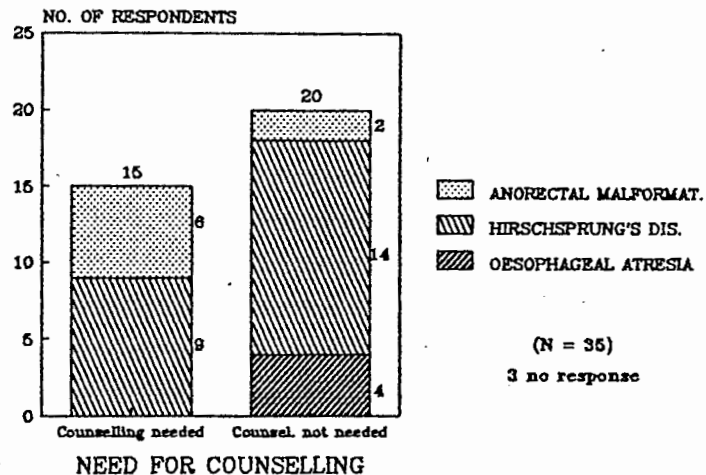


Diagram 16 indicates that OA respondents (4 out of 4) did not feel they had needed counselling concerning their anomaly, and more HD respondents (14 out of 23) felt they had not needed counselling, although 9 did feel that they had needed counselling. Most AM respondents (6 out of 8) felt that they had needed counselling concerning problems with their anomaly. Four HD respondents and 2 AM respondents had in fact received counselling at some stage of their lives.

Reasons counselling was needed - Depression (1 HD); sexual problem (1 HD, 1 AM); social embarrassment in public (1 HD, 1 AM); difficulties in mixing socially (1 HD); body image - scarring (2 HD); fertility problems (2 HD).

Additional comments by respondents concerning counselling - "... Now, I don't want to pity myself, but I think if I received counselling with regards to my illness at an earlier age (⁺ 12-15) I would have overcome some of the embarrassment I felt while growing up." (HD).

"... Baie meer sielkundige bemoeienis deur die hospitaal moet met pasiënte gemaak word." (HD)

Age at which counselling was needed - Depression (adult); difficulties in mixing socially (adolescence); body image (adolescence).

6.4 Demographic dataA. Sex/gender

Diagram 17 DIAGNOSIS BY SEX

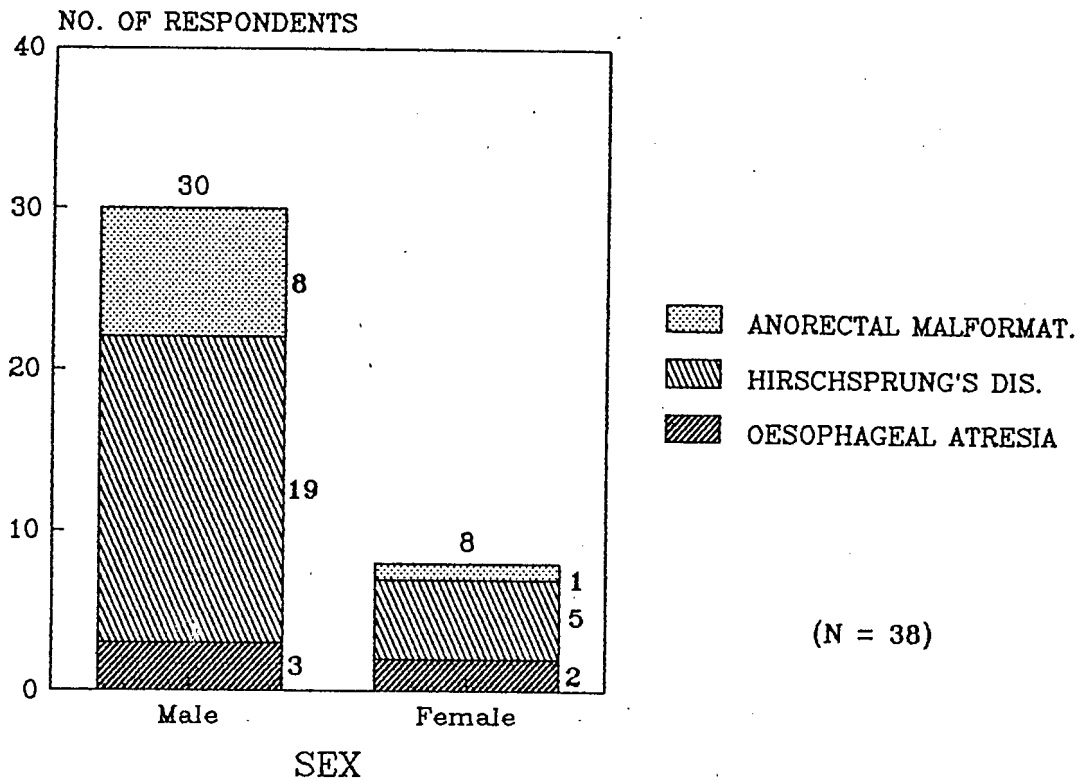


Diagram 17 reveals that there are more male OA respondents (3 out of 5), HD respondents (19 out of 24) and AM respondents (8 out of 9) than female respondents in each group, especially in the AM group.

B. Age

Diagram 18 DIAGNOSIS BY AGE

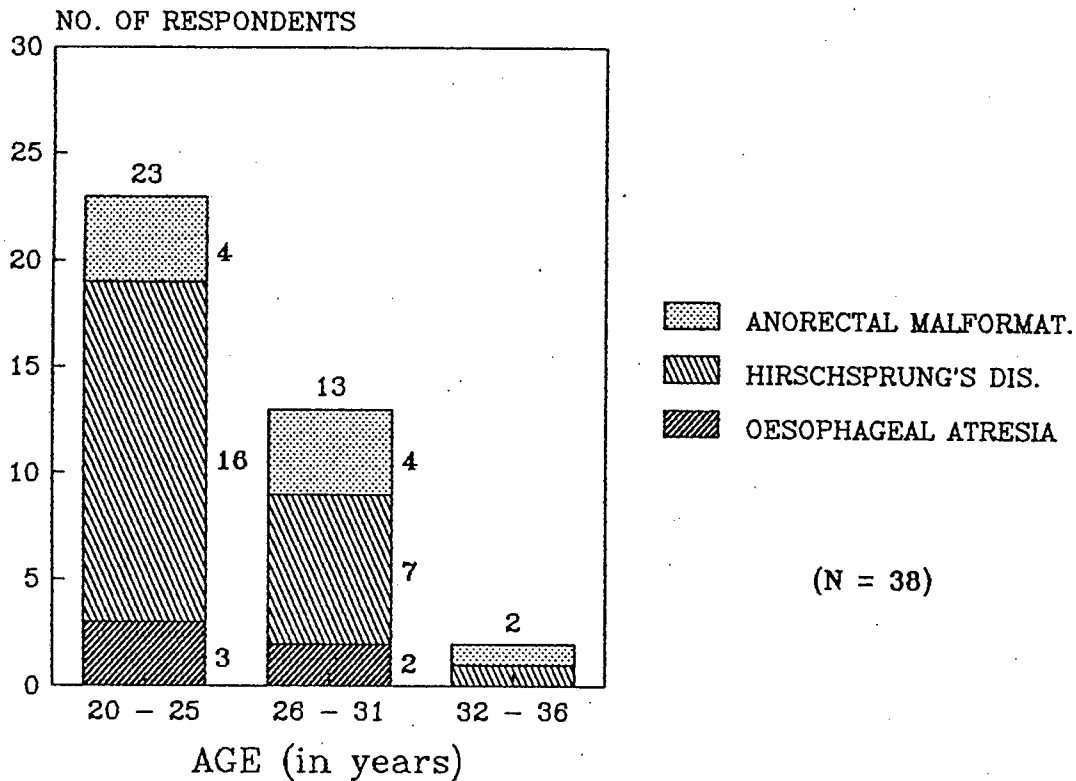


Diagram 18 shows that most OA (3 out of 5) and HD (16 out of 24) respondents are in the youngest age group, ie. 20-25, while AM respondents tend to be older and in the 26-31 and 32-36 age groups (5 out of 9).

C. Marital status

Diagram 19 DIAGNOSIS BY MARITAL STATUS

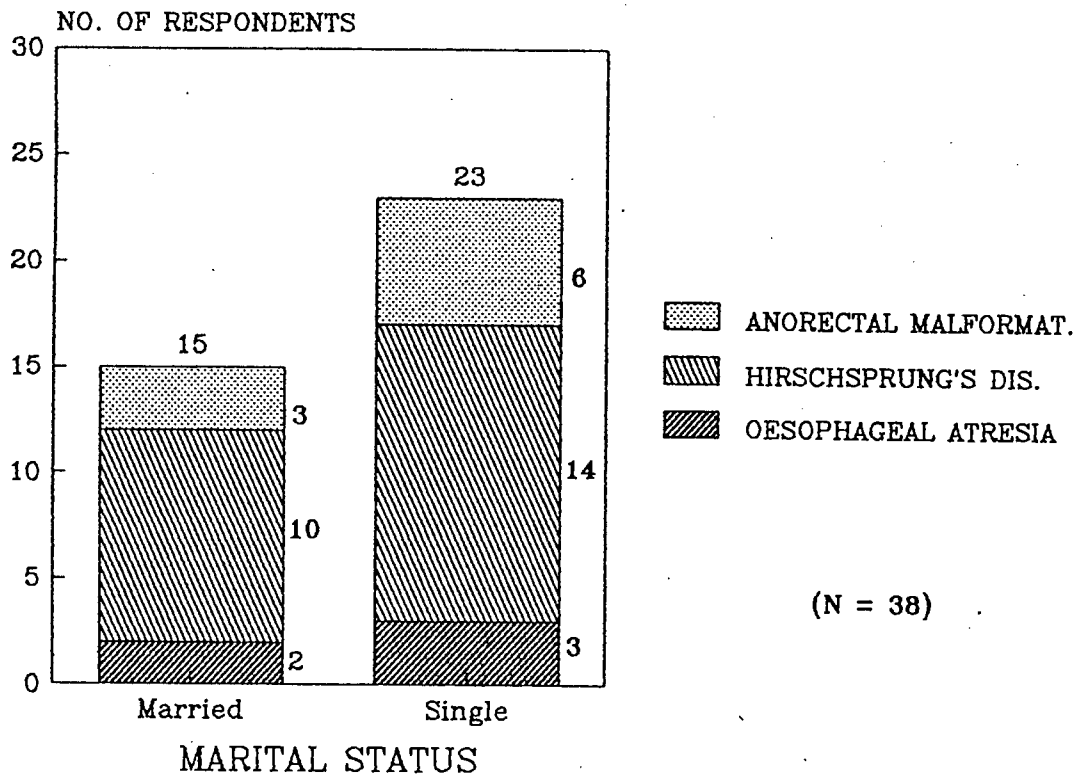
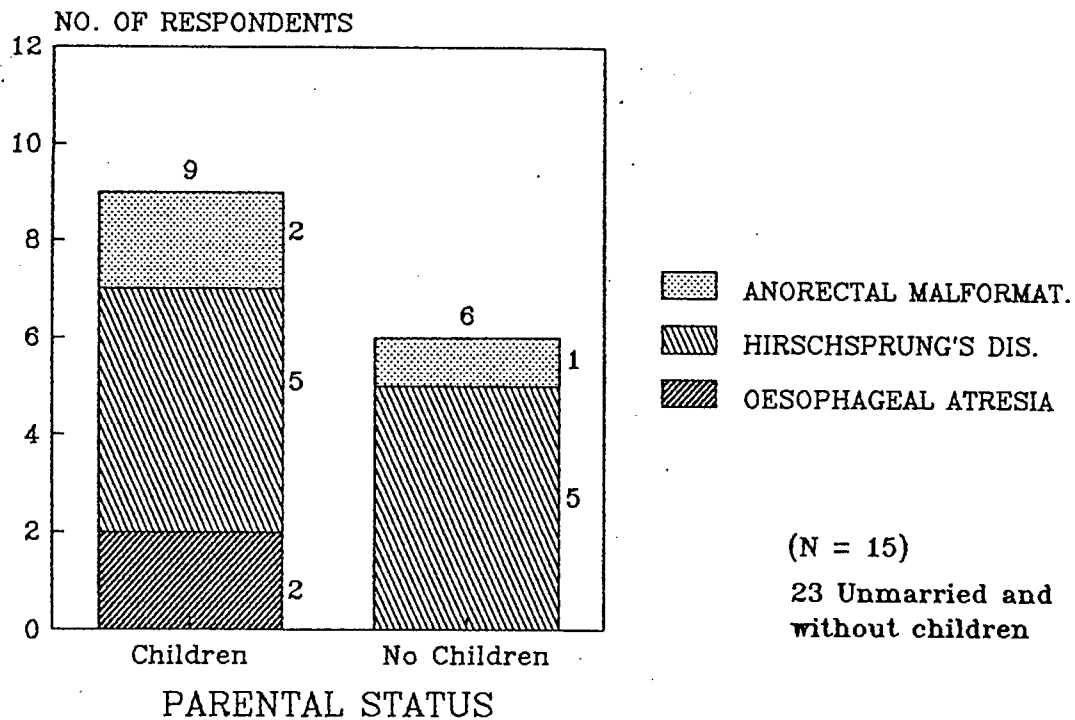


Diagram 19 reveals that more OA respondents (3 out of 5), HD respondents (14 out of 24) and AM respondents (6 out of 9) are single than married, with this trend being slightly greater in the AM group.

D. Parental status

Diagram 20 DIAGNOSIS BY PARENTAL STATUS *



* Married Respondent's Own Children

Diagram 20 indicates that all OA respondents who are married have children (2 out of 2). There is an equal distribution between HD respondents who have children (5 out of 10) and who do not have children (5 out of 10). More AM respondents who are married have children (2 out of 3) than those who are childless.

E. Religious attendance

Diagram 21 DIAGNOSIS BY RELIGIOUS ATTENDANCE

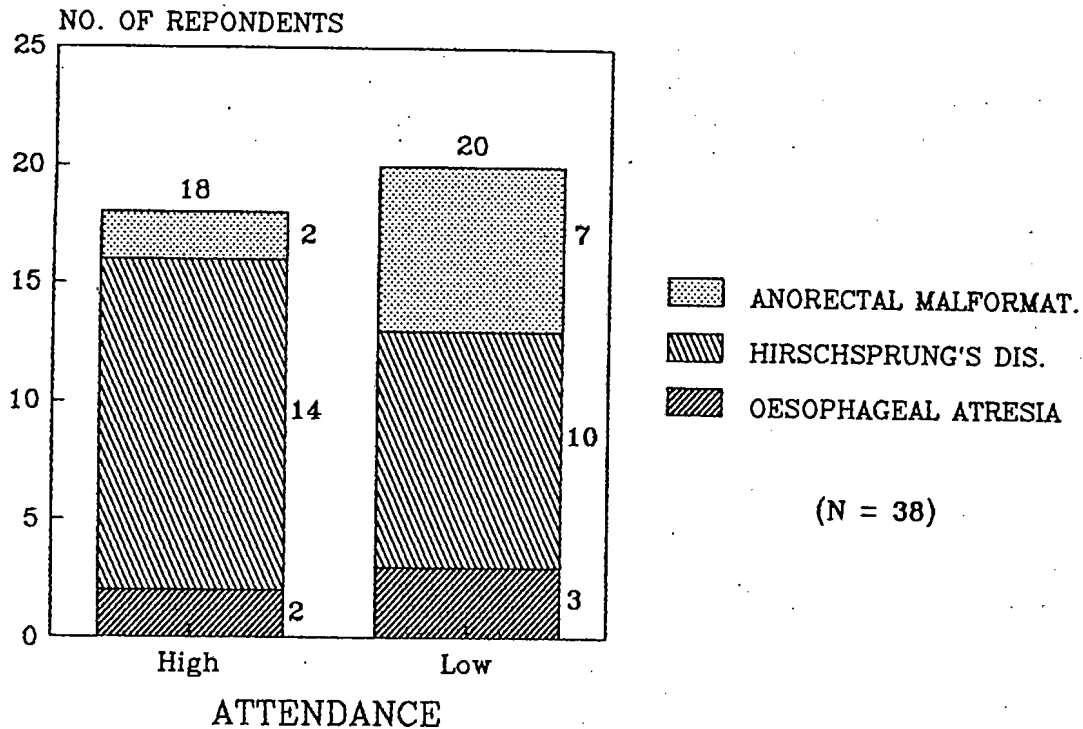
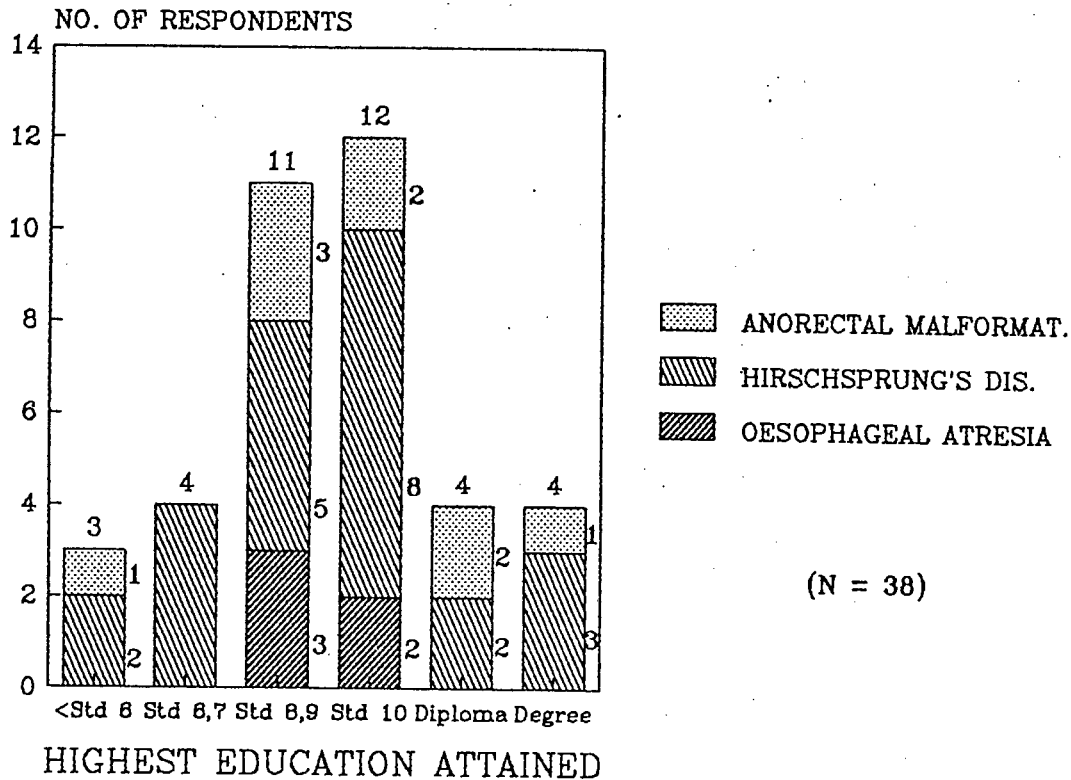


Diagram 21 suggests that OA respondents (3 out of 5) and AM respondents (7 out of 9) practise their religion infrequently, especially AM respondents, whereas HD respondents tend to practise more frequently (14 out of 24).

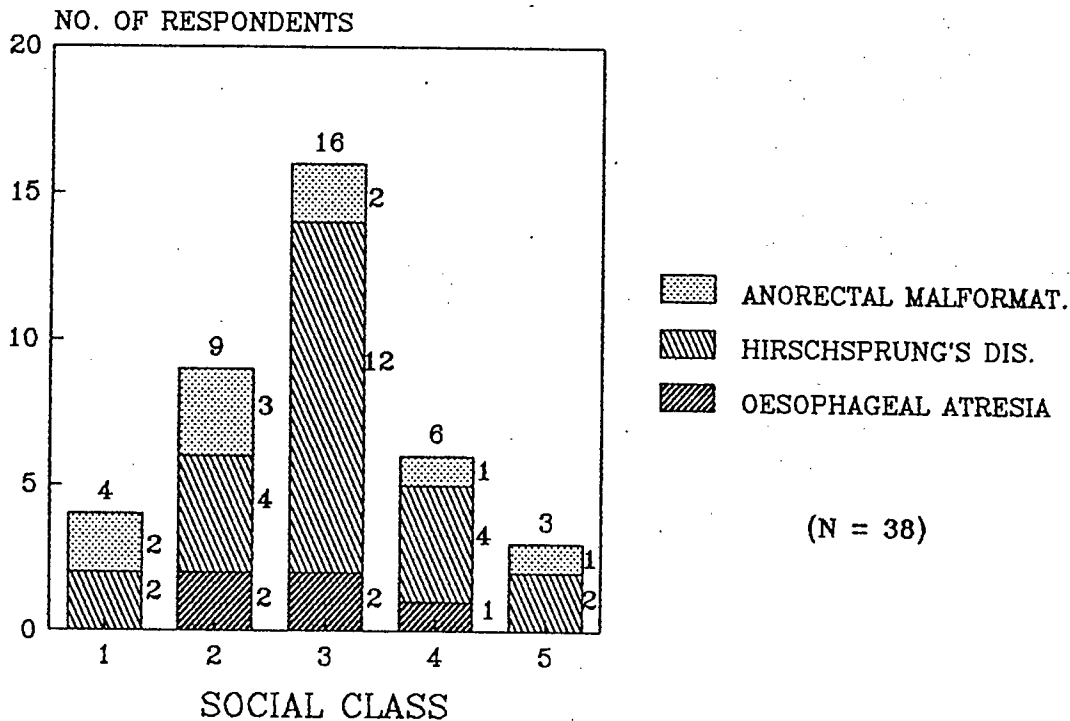
6.5 Socio-economic dataA. Education

Diagram 22 DIAGNOSIS BY EDUCATION



To simplify interpretation of this data education is classified in terms of respondents who completed and who did not complete school (primary and high school) and in terms of respondents who did or did not have post-school education.

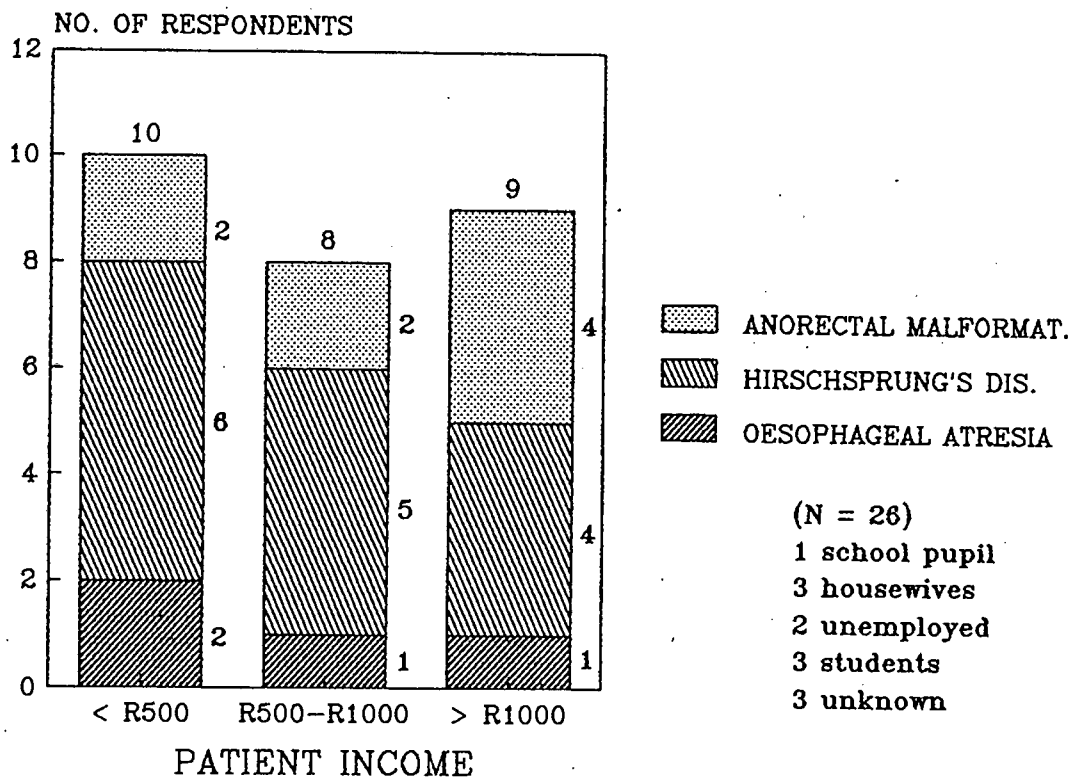
Diagram 22 indicates that more OA respondents (3 out of 5) have not completed school. None of these respondents have post-school education. There is a slightly higher tendency for HD respondents (13 out of 24) and AM respondents (5 out of 9) to have completed school with 5 and 3 respondents respectively having post-school education. Therefore OA respondents have lower education than HD and AM respondents. The latter have similar education, with AM respondents having attained higher education.

B. Occupational level (social class)Diagram 23 DIAGNOSIS BY SOCIAL CLASS
(OCCUPATION LEVEL)

Social class refers to the British classification based on occupational information (see Appendix 7). To simplify interpretation social class is classified in terms of social class I,II and social class III,IV,V. Diagram 23 reveals that there is a tendency for OA (3 out of 5) and HD respondents (18 out of 24) to be in social classes III,IV,V, while AM respondents tend to be in social classes I,II (5 out of 9). Therefore AM respondents are in occupations requiring more skills and higher qualifications than HD and OA respondents.

C. Income

Diagram 24 DIAGNOSIS BY INCOME



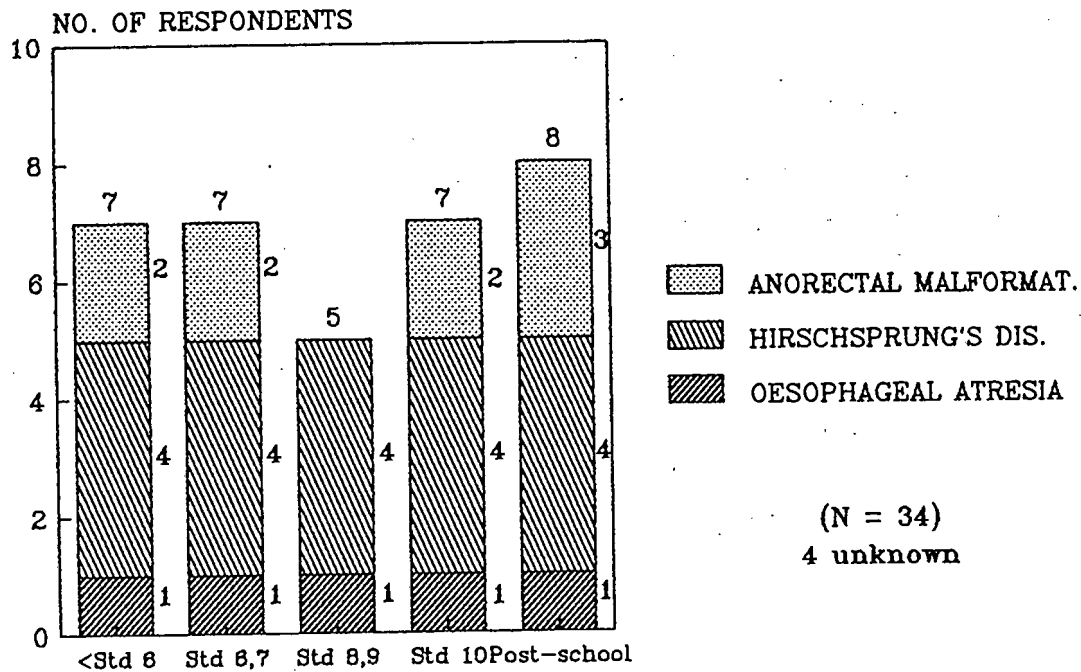
To simplify interpretation of this data, income is classified in terms of income below R500 per month and those with incomes of R500 and over.

Diagram 24 indicates that AM respondents tend to have more respondents in the R500 and over group (6 out of 8) and to a lesser extent the HD respondents also tend to have incomes of R500 and over (9 out of 15), whereas OA respondents have an equal distribution between respondents whose incomes are below R500 (2 out of 4) and R500 and over (2 out of 4). Therefore AM respondents have the highest incomes, followed by HD respondents, while OA respondents have the lowest incomes.

6.6 Socio-economic data - respondents' families

A. Education

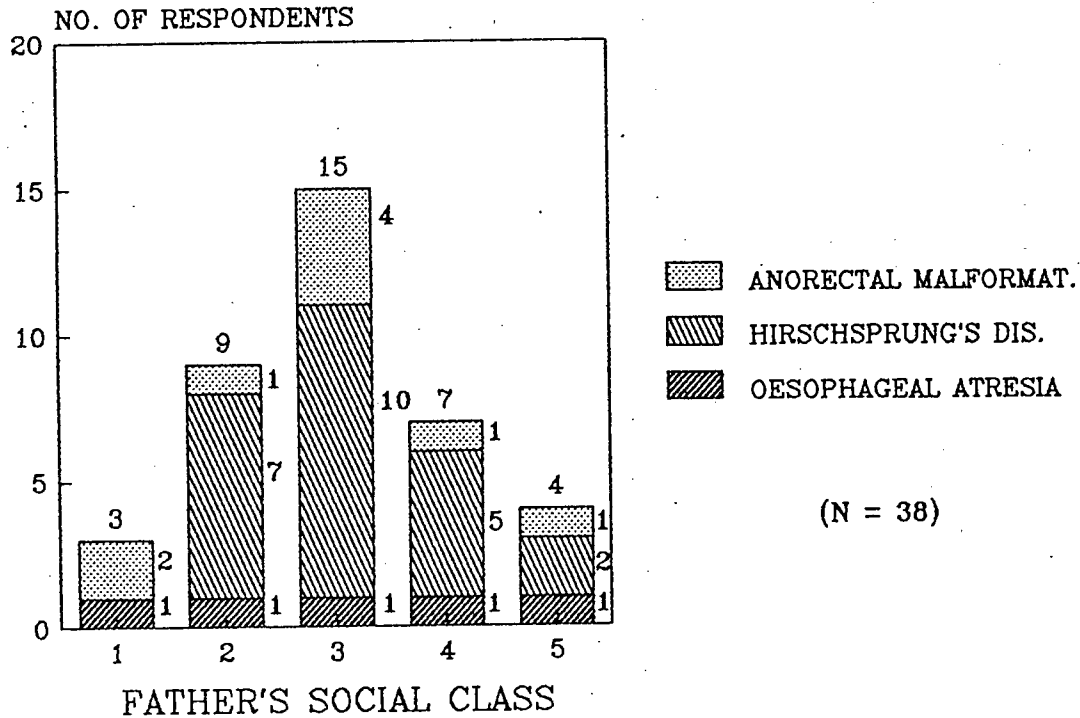
Diagram 25 DIAGNOSIS BY FATHER'S EDUCATION



HIGHEST EDUCATION ATTAINED

As the main breadwinner was usually the father in the present study (see Appendix 8), the socio-economic status of respondents' families was determined by the education and social class (occupation) of the father. There was no significant difference between maternal and paternal education. For the purposes of the study paternal education represents the education of the respondents' families (see Appendix 8 for maternal education).

Diagram 25 shows that there is a tendency for OA (3 out of 5) and HD (16 out of 24) respondents' fathers not to have completed school, whereas more AM respondents' fathers (5 out of 9) had completed school. The latter had proportionately more respondents' fathers who had post-school education (3 out of 9) than the HD (4 out of 24) and OA (1 out of 5) respondents' fathers. Therefore AM respondents' fathers have higher education than HD and OA respondents' fathers.

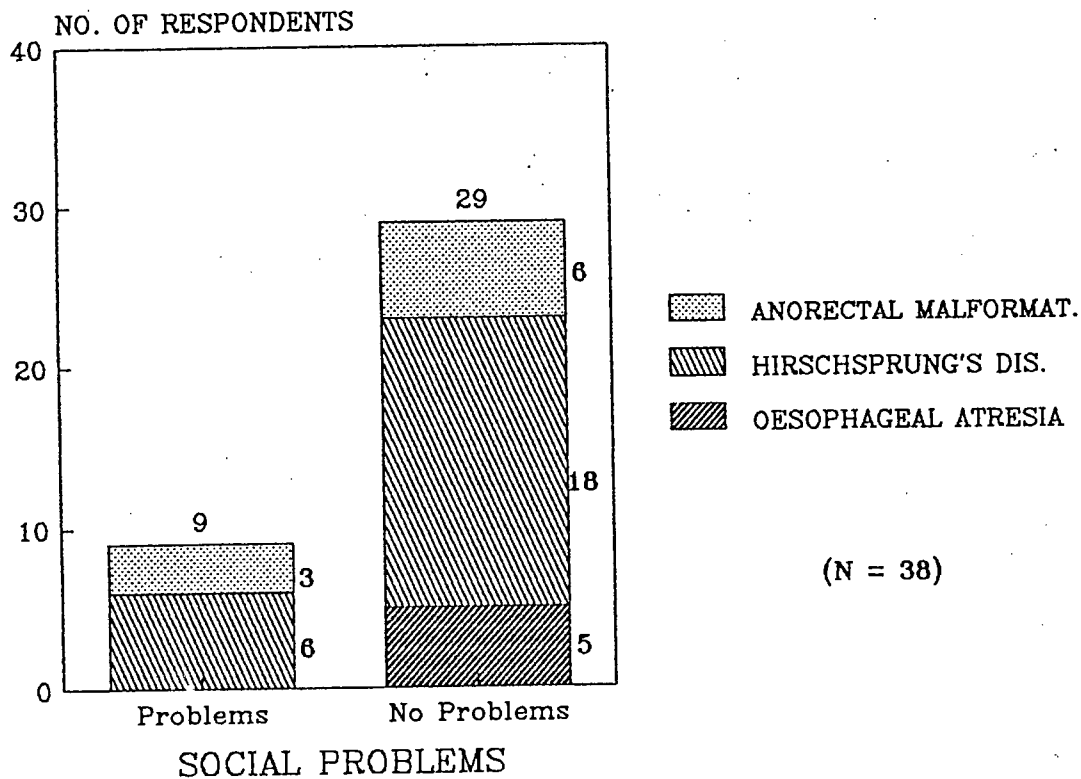
B. Occupational level (social class)Diagram 26 DIAGNOSIS BY FATHER'S
SOCIAL CLASS/OCCUPATION*

* Occupation represented by social class

Diagram 26 indicates that there is a tendency for OA (3 out of 5), HD (17 out of 24), and AM (6 out of 9) respondents' fathers to be in social classes III, IV, V. Therefore OA, HD and AM respondents' fathers are in occupations requiring similar skills and qualifications (see Appendix 7 for an explanation of the social class classification based on occupational information).

6.7 Family problem data (relating to respondents' families)

Diagram 27 DIAGNOSIS BY FAMILY PROBLEMS*



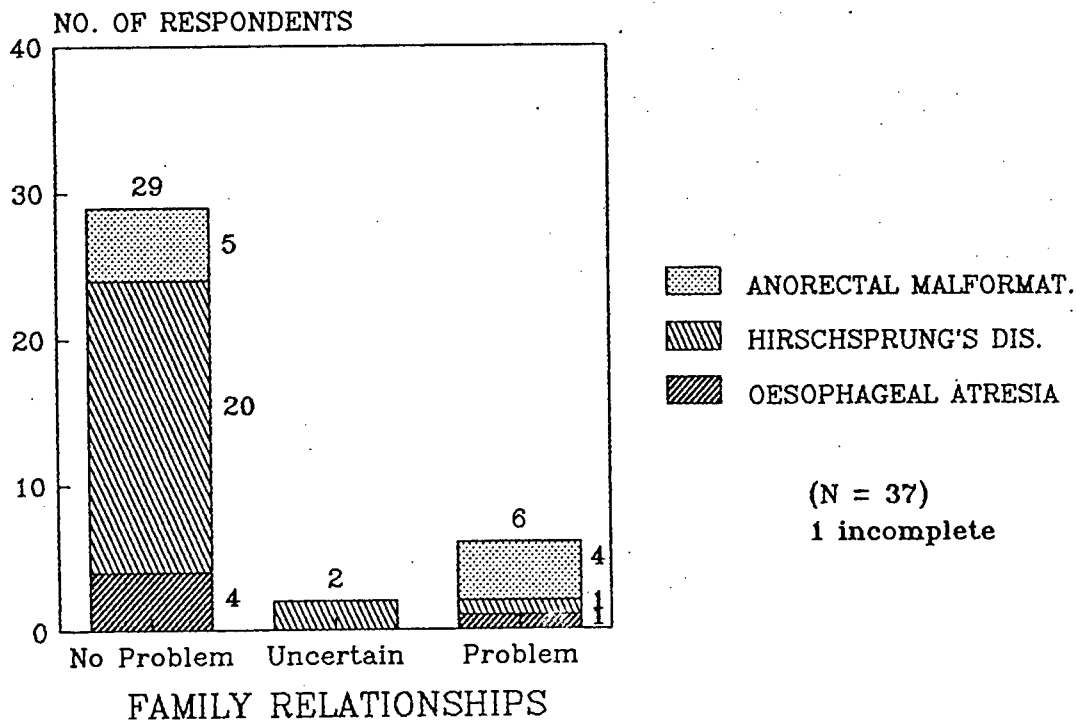
*Past problems in respondent's family

Diagram 27 shows that, according to respondents' reports, there are no OA respondents whose families experienced problems in the past and although more HD respondents' (18 out of 24) and AM respondents' (6 out of 9) families did not have problems, 6 of HD respondents' and 3 AM respondents' families manifested problems. Therefore AM respondents' families had the most problems followed by HD respondents' families, while OA respondents' families had no problems (see Appendix 8 for breakdown of family problems). (Family problems refer to problems in the past, while the respondent was growing up)

6.8 Psycho-social problem data (referring to respondents' current problems)

A. Family relationship problems

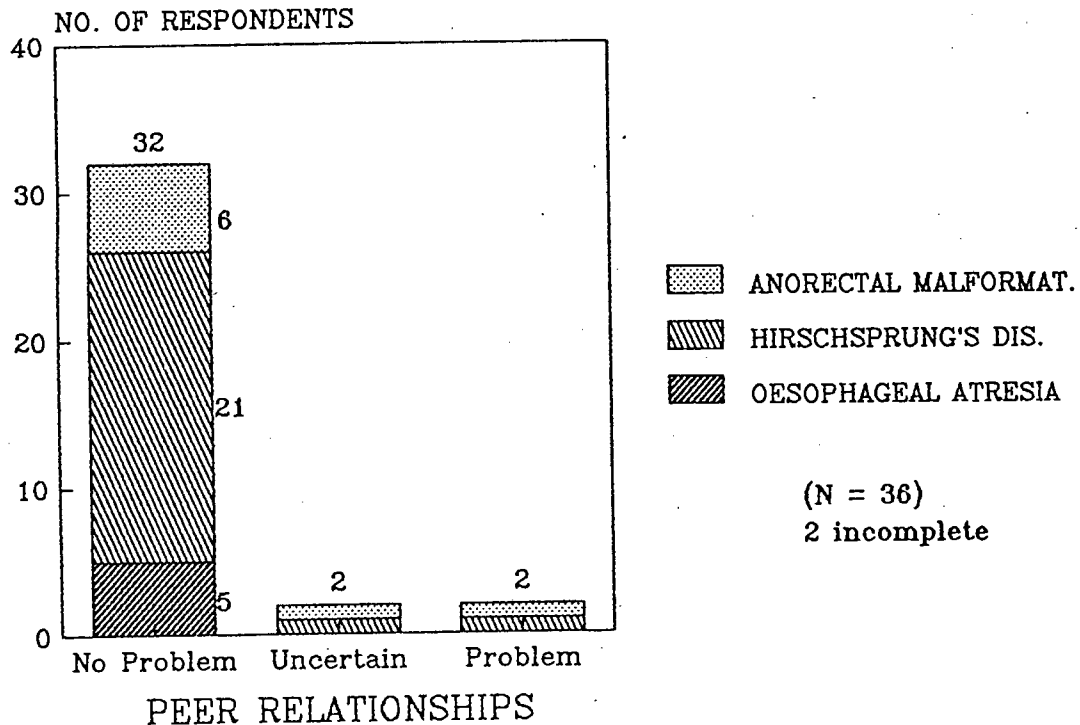
**Diagram 28 DIAGNOSIS BY PROBLEMS
WITH FAMILY RELATIONSHIPS ***



* Index of Family Relations

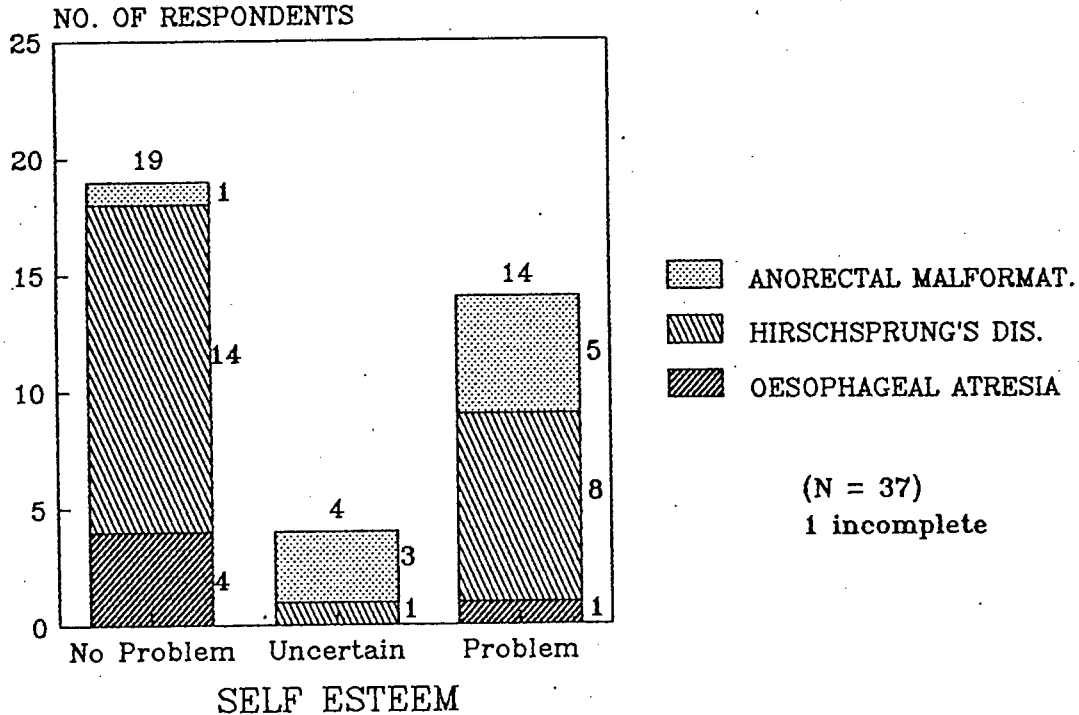
Diagram 28 indicates that, according to respondents' reports, there is a tendency for OA respondents (4 out of 5), HD respondents (20 out of 23) and AM respondents (5 out of 9) to have experienced no problems with family relationships, although 4 of the AM respondents do have problems. Therefore AM respondents have more problems with family relationships than OA or HD respondents.

(Family relationships refer to the respondents' present feelings concerning his/her family)

B. Peer relationship problemsDiagram 29 DIAGNOSIS BY PROBLEMS WITH
PEER RELATIONSHIPS *

* Index of Peer Relations

Diagram 29 reveals that, according to respondents' reports, most OA respondents (5 out of 5), HD respondents (21 out of 23) and AM respondents (6 out of 8) have no problems with peer relationships. AM respondents have proportionately slightly fewer respondents in the no problem group than OA or HD respondents.

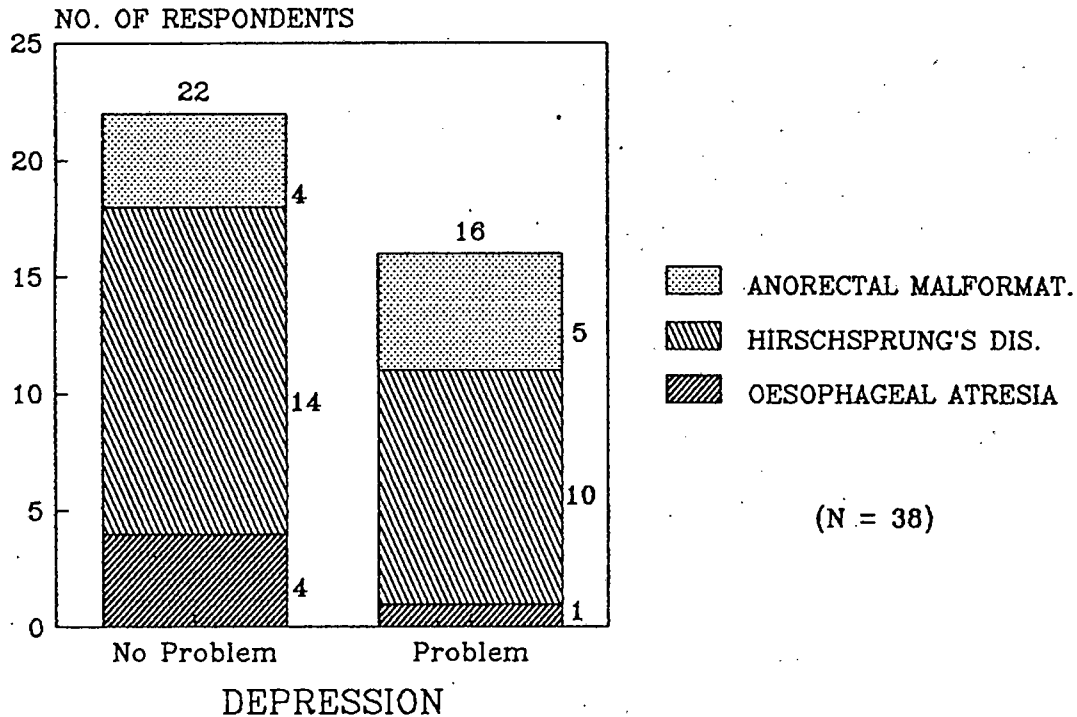
C. Self-esteem problemsDiagram 30 DIAGNOSIS BY PROBLEMS WITH
SELF-ESTEEM *

* Index of Self-Esteem

Diagram 30 indicates that according to respondents' reports there is a tendency for OA respondents (4 out of 5) and HD respondents (14 out of 24) to have no problems with self-esteem, although 8 of the HD respondents did have problems. There was a tendency for the AM respondents (5 out of 9) to have self-esteem problems. Therefore AM respondents experienced the most self-esteem problems, followed by HD respondents, with OA respondents having fewest self-esteem problems.

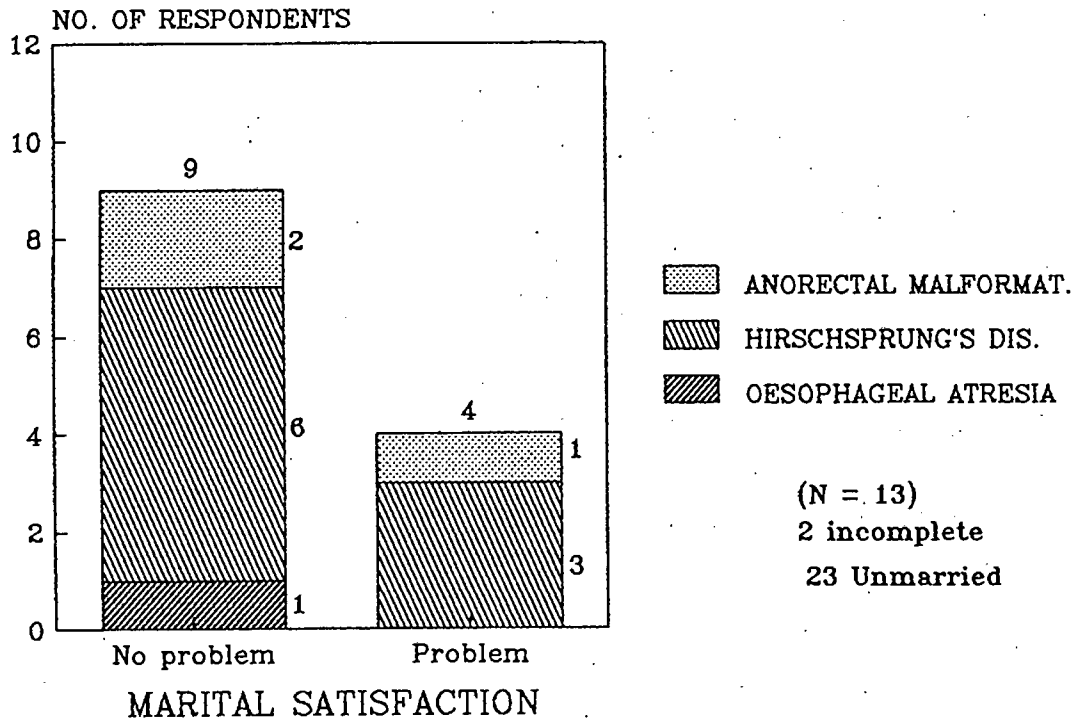
D. Depression problems

Diagram 31 DIAGNOSIS BY PROBLEMS WITH DEPRESSION *



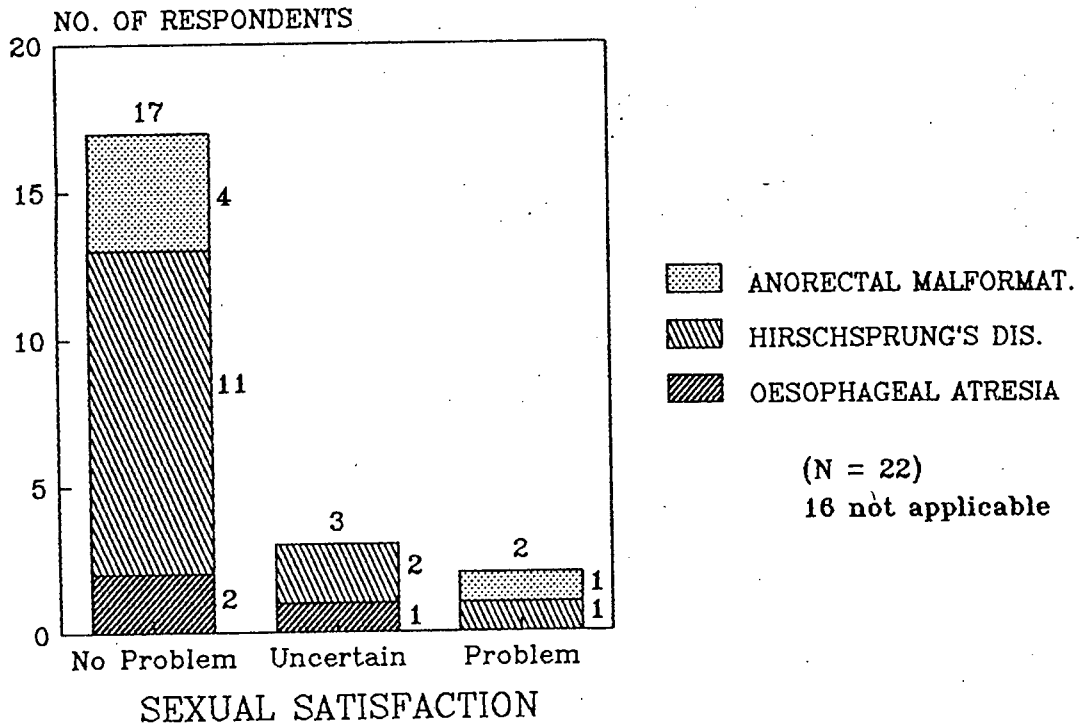
* Generalized Contentment Scale

Diagram 31 reveals that, according to respondents' reports, there is a tendency for OA respondents (4 out of 5) and HD respondents (14 out of 24) to have no problems with depression, although 10 HD respondents did have problems. There is a tendency for AM respondents (5 out of 9) to have depression problems. Therefore AM respondents have the most problems with depression, followed by HD respondents, with OA respondents having fewest problems with depression.

E. Marital satisfaction problemsDiagram 32 DIAGNOSIS BY PROBLEMS WITH
MARITAL SATISFACTION *

* Index of Marital Satisfaction

Diagram 32 shows that according to respondents' reports the OA respondents (1 out of 1), the HD respondents (6 out of 9) and the AM respondents (2 out of 3) experienced no problems with marital satisfaction, although 3 HD respondents did have problems. Therefore HD respondents have more problems than OA and AM respondents.

F. Sexual satisfaction problemsDiagram 33 DIAGNOSIS BY PROBLEMS WITH
SEXUAL SATISFACTION *

* Index of Sexual Satisfaction

Diagram 33 indicates that, according to respondents' reports, most OA respondents (2 out of 3), HD respondents (11 out of 14) and AM respondents (4 out of 5) have no problems with sexual satisfaction. (Sexual satisfaction problems referred to feelings about sexual relationships and not sexual functioning problems as reported in Diagram 10).

6.9 Information concerning the effect of the anomaly and residual problems on the respondent

A. Most embarrassing experience concerning congenital anomaly (Not all respondents answered this question. Some statements are quoted directly from the questionnaires, while others were made during the interview and are indirectly quoted). Research numbers prior to the quotes separate the responses.

(a) Oesophageal atresia respondents

OA 4 "... I once had to vomit due to food being stuck in my throat. I was in a restaurant at the time, I could not make the gents in time. At the same time I could not breathe. Food was stuck in my throat, coming up and going down again. I started to panic and eventually brought up on the carpet."

(b) Hirschsprung's disease respondents

HD 2 "... At a very young age I used to dirty my pants a lot, that is very embarrassing. When I am in company I can't help myself passing winds - it smells terribly."

HD 4 "... Eendag toe ek 15 was het ek met vriende gesels en het ek vuil geword."

HD 9 "... Om met die sakkie te loop en dit moes elke keer verwyder word om 'n ander een aan te sit."

HD 8 At a Scout camp at the age of 9 his nappy came loose and the other boys saw him and teased him. He never went back to Scouts.

HD 20 "... Geen beheer oor opelyf en winde."

HD 21 "... Having no control over your bowels when at primary and secondary school. Smearing in pants during present married life."

HD 23 "... Om die eerste keer my littekens aan my eggenoot te ontbloot."

HD 10"... Miskien toe ek in my kinderjare my broek vuilgemaak het.

Dit het my skaam gemaak as my moeder moes besluit of ek mag uitgaan of nie want dit hang af of daar 'n toilet is of nie."

HD 12"... Several occasions when my tummy rumbled unexpectedly. When with girls/girlfriends when they did not know about my illness - mainly in my teens. This still happens today, but is accepted readily."

HD15 "... Merke op my broeke: Kinders wat spot."

HD17 Had an accident in his pants on the train once which was embarrassing as everyone was looking at him.

(c) Anorectal malformation respondents

AM 2 "... Having no control over my bowels one day while in class at primary school."

AM 3 When playing rugby he was tackled and his colostomy bag broke and the contents ran down his legs which was very embarrassing.

AM 4 "...The looks you get when you break a wind."

AM 8 "...Op kosskool het ek my bed op twaalfjarige ouderdom natgemaak."

AM 9 "...Op skool in Std 2 tydens 'n liggaamsoefening klas was daar 'n merk aan my broek. Een van my klasmaats het dit gesien en my daaroor gespot. Gebeurtenisse soos hierdie het my selfbeeld 'n ontsettende knou gegee."

B. Effect of operation and residual problems on respondent's life (Not all respondents answered this question)

(a) Oesophageal atresia respondents

OA 14"... Never affected my life."

OA 2 "... I still have abnormal coughing and occasional choking. My throat makes strange noises and I get an occasional closed feeling in my throat."

OA 3 "... Voice fluctuates uncontrollably."

OA 4 "... It has affected my life in that I could not take part in life in a completely normal way. But it is not really so restricting except for the odd throat ailment."

(b) Hirschsprung's disease respondents

HD 10 "... Nee, die operasies het my lewe geensins beïnvloed nie."

HD 15 "... Vind dit moeilik om te kommunikeer. Sou nie in die huwelik tree as gevolg van seksuele probleem."

HD 12 "... Since pouch operation my quality of life has improved as I can now do things I could not previously."

HD 9 "... Ek kan nie kinders kry nie."

HD 3 Incontinence throughout his life affected the patient so negatively that at the age of 20 he tried to commit suicide.

HD 9 "... Op die ouderdom van 6 jaar het ek skool bygewoon en later moes ek met die sakkie loop en het ek baie skaam gevoel. Dit was later verwyder en het ek veel beter geword."

HD 8 This affected his confidence when he was young. He did not mix with others, only his brothers and sisters.

HD 5 "... Grew up with a lot of embarrassment concerning scars. I also found it embarrassing to go to the beach and have to explain my scars to people. After a while I took to wearing a sweater on the beach and keeping it on, even while swimming. When I started primary school I still had a hole in my abdomen, which leaked when I exerted myself physically. After having to answer numerous questions from the teachers and pupils alike, like why I am dirtying my vest and not my underpants, I quietly withdrew

from physical activities at school. It was only during my matric school year and while at university that I started getting involved in sports again by playing rugby. I sometimes still don't want to take off my clothes when I go to the beach, especially when it's crowded."

HD 17 Problems in childhood, eg. no control over bladder which he was teased about at school, caused him to stutter which has continued to the present time.

HD 20 "... Kan nie aan enige sport deelneem nie"

HD 21 "... With only a few social and psychological problems and a scar on lower abdomen, no other after-effects restricted my life."

(c) Anorectal malformation respondents

AM 5 Problems in childhood concerning his residual problems affected his confidence negatively.

AM 8 "... Partykeer in die verleentheid oor winde, ens., en die feit dat ek geen kinders kon hê nie was in die begin moeilik om te aanvaar."

AM 1 "... Wel, ek kan bly wees ek leef, gesond is en afgesien van my probleem 'n man met 'n familie is."

AM 2 "... Apart from the fact that I soil my pants fairly regularly and drip a bit, particularly after exertion, I find my problem OK to cope with."

AM 4 "... Made me choose my diets more carefully and to take things as they come."

AM 9 "... Ek is selfbewus oor die gedeelte van die rektum wat uitsteek, asook oor die ander probleme, bv. merke aan my broek."

C. Additional comments on medical questionnaire

(a) Oesophageal atresia respondents

OA 2 "... I feel I have adapted to my throat problems and sounds it makes and have learnt which foods make it worse, ie. dairy and cereal products."

(b) Hirschsprung's disease respondents

HD 15 This respondent was very depressed about being infertile. He said that he did not feel like a man, even though it did not worry his wife. He was also embarrassed about smearing marks on his pants and felt that as the man of the house this was very humiliating to have. He said he did not want to give his pants to his wife to wash, but she insisted on washing them. He said if he had known before his marriage that he could not give someone children, he would never have married.

HD 17 "... Ek is ook skaam om kaal bo!f te loop want die merke is nog steeds duidelik sigbaar."

This respondent is worried about getting married and his wife seeing the marks on his pants. He said he is a man now and should not have that problem.

HD 3 When he was in primary school no-one would play with him, because he smelled. So he paid children to be his friend for the day so that he had someone to play with. Often he would just sit at breaks and cry as no-one wanted to play with him. When he got older he found that having no control over his bowels meant not being able to get a girlfriend. As soon as a girl found out she would leave him. He used to go to dances and sometimes would have an accident in his pants and then could not get up for the rest of the evening to dance. Eventually he was so depressed he tried to commit suicide at the age of 20.

HD 8 This respondent said that the most important thing is to keep your problems a secret and not let anyone know. When he was young he used to only go out with his brothers and sisters who would cover up for him if he had an accident. He had to think carefully before going to places because if there was not a toilet, he could not go. He sometimes found it difficult to hide as he had to wash his own nappies and he was scared that people would see him hanging up his nappies when he was ten and they would wonder what he was doing. He said that it must be hidden from everyone and that even his wife does not know about his problem.

(c) Anorectal malformation respondents

AM 7 The respondent said that he could not get a steady girlfriend as he was scared they would discover he is still soiling and so he never allows anyone to become too close to him. He finds it very difficult to mix socially which is further aggravated by the fact that he can't drink alcohol as it gives him diarrhoea.

AM 2 "...I find you learn to tolerate the situation because in my case I don't think it's such a big deal."

AM 9 "... Die mense wat weet van my probleem, is onder die indruk dat ek dit baie goed hanteer (bv. my ouers). Ek is egter 'n introvert en hou alles vir myself en probeer om nie my ware gevoelens te wys nie. Geeneen van my vriende weet van my probleem nie. As gevolg van my probleem, dink ek het ek nie genoeg selfvertroue opgebou nie. Ek het my soort van onttrek van meeste sosiale funksies. By die skool, het ek 'n bietjie tennis en krieket gespeel maar kon byvoorbeeld nooit saam met maats op ander plekke aan sport gaan deelneem nie."

(d) All anomaly groups

Many respondents in all groups referred to having residual problems, but feeling 'too old' for The Children's Hospital. Those who attended an adult hospital often reported feeling unknown and strange.

6.10 Case studies - description of information received during interviews

(one case study from each anomaly group)

The following information concerns subjective reports from 3 respondents (10A, 1HD, 1AM) which were collected during in-depth interviews. The reports are in response to questions concerning the effect of residual problems on various parts of their lives. The effect of residual problems was examined in the past (aspects of respondents' lives up to the age of 19) and in the present (aspects of respondents' lives from the age of 20 onwards). The information given was in response to closed and open-ended questions (see Appendix for the semi-structured interview schedule - Questionnaire D).

A Oesophageal atresia (OA 4)

Description of respondent

The respondent was male, 26 years, single, from social class III and was still experiencing problems with swallowing on a daily basis and problems with vomiting about twice a year.

(a) Effect of residual problems on aspects of respondent's life

(Effects in the past)

(i) Hospitalization

The respondent could not remember much about his hospital experience. He knows he had one operation and feels that the experience in hospital had no effect on his life.

(ii) Family

He describes the effect on his family as minimal, except that his parents would prefer not to eat out at friends or restaurants because of his swallowing and choking problems while eating.

(iii) School

He describes the effect of residual problems on his

schooling as minimal. He was absent about once every six months because of hospital appointments. Residual problems did not affect his schoolwork in any way. He failed standards one, four and ten, but does not relate this to any medical problems. He had a few embarrassing incidents at school - vomiting in class once and twice on the playground - but he did not see this as a big problem.

(iv) Friends

He had many friends (8-9), had no difficulty in making friends and in no way did his residual problems affect his relationship with his friends.

(v) Recreation/social life

In no way did residual problems affect sport and recreational activities or his social life.

(vi) Dating

The respondent's residual problems had no effect on his dating when he was a teenager or his sexual relationships.

(vii) Mental health

While he was depressed, anxious and used alcohol occasionally, he did not feel that this was related to his residual problems. He had no sleeping difficulties, rarely smoked cigarettes and had no weight problem. In his opinion nothing upset him about his residual problems.

(b) Effect of residual problems on aspects of respondent's life

(Effect at present)

(i) Post-school education and occupational information

The respondent is a printer and his illness did not affect the choice he made in studying his course or his choice of occupation. He works full-time and has not had a problem

with unemployment, absences from work, problems with those he worked with related to his residual problems or the way he did his job. This also applied to a previous job as a packer in a biscuit factory. He completed his military service and this was not affected by his residual problems.

(ii) Peer relationships

The respondent has 6-10 close friends, had seen a close friend in the previous week, experienced no difficulty in making friends, experienced no embarrassing incidents at present related to residual problems, did not see these problems as affecting his relationship with friends and had told all his friends about his anomaly.

(iii) Marital

Not applicable - unmarried.

(iv) Dating

The respondent does go out on dates, but has not had steady relationships. Residual problems have not affected his dating or relationships and if he does experience problems he can discuss them with his partner.

(v) Sexual relationships

Although he is not having a sexual relationship at present, he has had relationships since his teenage years. He does not see residual problems affecting these relationships or his sexual functioning or how sexually attractive he is to others.

(vi) Mental health

He feels satisfied with life as a whole, satisfied with himself in general, very satisfied with his appearance and the part of his body he dislikes the most is his left foot

which is slightly abnormal in appearance (unrelated to his anomaly). He occasionally feels depressed, rarely feels anxious, rarely uses dagga, occasionally eats less than usual when upset, likes mixing with people, has never served a jail sentence and he feels that he has no psychological difficulties at present. While having difficulty falling asleep, using alcohol and cigarettes often, this is not related to his residual problems. He again mentioned that nothing upsets him at present about his residual problems.

(vii) Recreation/social life

He rates his social life as good and cycles and swims in his spare time. He participates in sport and belongs to two organizations. He enjoys being with others in his spare time and can take part in all activities that they do and in no way is his social life, sport or recreational activities affected by his residual problems.

(c) Medical and Social Work Services

(i) Medical services (past and present)

He could not remember a great deal about his experiences at the Children's Hospital. He feels he received excellent treatment (medical and personal) at the hospital and that his operation was successful. He was first told about his operation when he was at primary school age by his parents. He came back to the hospital after the age of 14 once and did not mind returning to the Children's Hospital. He later went to Groote Schuur Hospital for dilatations - this transition did not bother him. He does not think there is a need for a clinic specifically for teenagers born with his anomaly, but suggested that the adult hospitals be divided up into different wards for different age groups. He had

no suggestions for improving medical services at the Children's Hospital.

(ii) Social Work Services (past and present)

He has not received any professional counselling and does not feel he needed it or that people born with his anomaly need it.

(d) Additional comment

He was very thin at school and is not sure if this was related to his anomaly - but it was not a big problem.

(e) Evaluation of interview

He did not mind answering questions at all.

B. Hirschsprung's disease (HD 23)

Description of respondent

The respondent was female, 24 years, married, from social class III and was presently experiencing problems with scarring on her abdomen, abdominal distension and infertility. In the past she had frequently been ill with problems relating to her anomaly.

(a) Effect of residual problems on aspects of respondent's life

(Effect in the past)

(i) Hospitalization

She had spent a lot of time in hospital and had had five operations. She was diagnosed very late (at the age of six) and had spent one year at the age of 6, three months at the age of 10, three months at the age of 15, one month at the age of 16, two months at the age of 18 and ten days at the age of 23 in hospital. She felt that the time she spent in hospital had had a negative effect on her life. When in hospital the first time she felt that she could not ask for some things she wanted as she felt like a burden on the staff because she was there so long. She also knew that her parents were not paying very much so she

felt that she could not ask for anything extra. She spent a birthday, one Christmas and one Easter in the hospital which upset her. She missed her family a great deal and when she came back home she felt like a stranger as she had missed out on so many family activities. She also felt that she hardly knew her brothers and sisters as they were not allowed to visit her in hospital. Her parents used to pretend that they were going to the toilet when they had to leave after visiting her because they did not want her to cry, but she looked out of the window and saw that they were leaving - this upset her a great deal. She also had a fear of dead people, as many children died and were put in a special room, and the children were scared of it. She said that although she made friends, they did not usually stay for long.

(ii) Family

The residual problems and being ill for a large part of her childhood had a very disruptive effect on her family. It restricted family activities and affected the relationship with her brothers and sisters. When she was ill her mother could not sleep, there was an unpleasant atmosphere, her sisters used to cry and her brothers and sisters could not study as everyone was worrying and fussing around her. Often her illness restricted family activities. For example, if there was going to be a party at the house and she was sick, the party was cancelled. She felt like a 'spoilsport', as if it were her fault. She was overprotected as a child. If she hurt herself while playing, her brothers and sisters received hidings. She was always the centre of attention in a way she did not like. For example everyone in church knew about her illness and when she finally returned to

church, they were very sympathetic, but never treated her like a normal person. When being introduced to new people, her family would tell them about her sickness, and she would have to show them her scars. She had a very poor relationship with her mother, as she blamed her mother up to the age of 18 for her illness as her mother only took her to the hospital when she was six. Her father left the family when she was young.

(iii) School

Her illness had a negative effect on her schoolwork, as she was frequently absent and found it difficult to catch up the work. She was absent from school about four times each year for check-ups and was frequently absent because of illness or periods spent in hospital. She failed matric because of spending so much time in hospital. She had many embarrassing incidents at school concerning her scarring and abdominal distension. She was always worried during PT that her shirt would slip up and that someone would see her scars. Apart from her scarring she also had bowel problems, mainly constipation and abdominal pain. It was not really an obvious sort of problem and the teachers thought she was just being lazy when she did not want to do exercises. In high school she found it embarrassing to tell male teachers about her problem. At school they tried to get her involved in sport and because she was thin, they thought she was fit and healthy, and would ask her "Why don't you do long jump, you've got such long legs?" She had a problem with her abdomen distending and could not do up her school belt and other children teased her about this. She used to wear a corset in high school to try and make her slimmer around her abdomen. She never changed in front

of the other children and would slip out the period before PT and put on her PT clothes under her school clothes so that no-one would see her scars.

(iv) Friends

She had two close friends when she was a child and as a teenager. She did not have difficulty in making friends and did not think that her problems affected her friendships. She did not tell her friends everything about her illness and when they came to visit her when she was sick, she used to wear a few pairs of pants so that they would not see the ridges that her scars made on her abdomen. It was too embarrassing to go to friends' toilets as it used to smell so badly, so she would not go and hold it in until she returned home - this was very uncomfortable..

(v) Recreation/social life

She found that her residual problems affected her sport and recreational activities a great deal. She could not take part in ballet because of her large abdomen. She could not go mountain climbing as she got too tired. When she went to a party everyone would check to see whether she could eat certain things. Swimming was always a problem as the scars would show through her bathing costume when it got wet. Also other people would ask her "Why don't you wear a bikini, you're so thin?" As soon as she climbed out of a pool or the sea, she would put on a robe in case anyone saw the scars and when she lay down in the sun, she would put a towel over her stomach. Although her problems prevented her from going away on holiday, they did not prevent her from staying overnight at friends. Another embarrassing thing was that her stomach made noises in public and she knew everyone knew it was her stomach.

(vi) Dating

She began dating at the age of sixteen. She had four dates when she was a teenager and a steady relationship. Her residual problems had a large effect on her dating and relationships. She could often not go out on dates because she was frequently ill. She did not want boys to visit when she was ill as she found it embarrassing to tell them that she was constipated. She did not want any boys to get too close to her in case they heard the noises that her stomach was making. She also did not want boys to touch her stomach and feel how big it was. When she went to ballroom dancing, she was scared that a boy would put his hand on her stomach and feel how big it was or feel the scars. She did not have any sexual relationships when she was a teenager, but this was not related to her residual problems. However, she feels that she would not have had any because she was worried about being naked in front of anyone because of her scars. When she was a teenager and talking with her friends, they used to talk about getting married and she was convinced that she would never get married because of her scars so these conversations used to upset her.

(vii) Mental health

She was very often depressed and anxious because of her residual problems. She was satisfied with her overall appearance, did not have difficulty sleeping, did not use alcohol or smoke cigarettes. She was very thin when she was young. Her constipation and scarring were the aspects concerning her residual problems that upset her the most. She said that it turns your whole life upside down. At the age of sixteen she had a nervous breakdown and cites being so anxious as the reason for

this.

(b) Effect of residual problems on aspects of respondent's life

(Effect at present)

(i) Post-school education and occupational information

The respondent is presently a housewife and does part-time dressmaking. She would have liked to study, but after having to repeat matric because of illness, she was worried that this might occur again. (Her brothers and sisters all studied further). She wanted to be a model but she knew she could not because of her large stomach. She had worked previously for a clothing company, but was often absent. She prefers to work at home now as she is tired of telling people about her illness.

(ii) Peer relationships

She has about three close friends, one of whom she had seen on the previous day and has no difficulty making friends. Her problem does not affect her relationships and her close friends know about her illness.

(iii) Marital relationship

This is her first marriage. She married an understanding man who accepted her problems, eg. being sick often. She feels her residual problems are affecting her marriage. She often gets ill and feels like a burden on her husband. If she feels ill she does not want sex and she feels bad about this. This used to occur a lot in the early days of her marriage. Her husband knows about the illness, but she does not feel that she can talk easily to him about problems - she feels that her marriage is good, but that honesty and communication are poor. She feels that her marriage would have been different if she had not had scarring and problems with sex.

(iv) Sexual relationship

She views this relationship as poor. She does not feel sexually attractive because of her scars. She finds that the pain affects her functioning and she suffers from vaginismus, but not orgasmic problems. She feels that her sexual relationship would have been different if she had not had any residual problems. For example, because of her scarring, she is not keen to try certain sexual positions and she still does not like her husband to see her naked, see her scars, or to put his hand on her stomach. Apparently this does not worry him, but it worries her. One of the most upsetting problems that she has, related to her anomaly, is that she is infertile. According to various gynaecologists it seems unlikely that she will have children because of her extensive internal scarring. She has not told her husband's parents yet and they frequently ask why she does not start having children. Although this upsets her husband he has been very understanding. She is very worried that her in-laws will be furious about her infertility, as unless she has children his side of the family will not continue, as there is no one else to have children. She feels sometimes that her husband should get another woman who can give him children. She does not want to adopt a child as she has seen adoptions that have not been successful.

(v) Mental health

She is satisfied with life as a whole, with herself and her appearance in general. Her stomach is the part of her body she dislikes most and said ' ... I used to feel that I would rather be blind than have this stomach. I felt that I would want any other part of my body messed up, but not this part - not my

stomach.' She felt that it was unfair that it was her stomach that was so unattractive when she looked at other people with disabilities. She said ' ... You think your problem is the worst and that no-one else has a problem as bad as yours.' The problem was worse in a way in that she looked normal with her clothes on. She used to think, when men found her attractive, that if they could see behind her clothes they would get a big surprise. She occasionally feels depressed but rarely feels anxious, rarely has sleeping difficulties and never smokes or uses dagga or alcohol. Her eating is not affected when she is upset. She has never served a jail sentence. She likes mixing with people. She does not think she has any psychological difficulties at present. Not being able to have children is the aspect about her residual problems which upsets her the most at present.

(vi) Recreation/social life

She feels her social life is good. She spends most of her spare time sewing. She cannot do all the sports she wants to do because of pain in her abdomen. For example, her friends attend keep-fit classes and she cannot go.

(c) Medical and Social Work Services

(i) Medical Services

She feels she received good treatment at this hospital overall and that her operations were successful. However, she experienced some problems with nursing treatment. She said that the nurses did not always look after her properly and when the doctors asked about what had been done, they did not always tell the truth. For example, they did not always change her bed immediately if she wet it, and did not always change her

colostomy bag if it needed it. If she reported it, they used to shout at her so she finally said nothing. She did not feel that the doctors gave her enough information concerning her anomaly. She had positive contact with the hospital as she liked her surgeon and he made all the difference. After the age of fourteen, she went to Groote Schuur Hospital. She hated this change and would have preferred to return to the Children's Hospital. She found the new hospital strange and frightening. She suggested that there should be a ward for teenagers attached to adult hospitals.

(ii) Social Work Services

She has received professional counselling from a psychiatrist. She feels that she needed this type of help a few times in her life, but mostly when she was eighteen. At this stage she was very worried about marriage and fertility, as she realized she might be infertile. She feels that people born with her anomaly do need counselling - especially about how their residual problems will affect their future. She has not received any social work services from the Children's Hospital. She suggested that follow-up studies, like the one under discussion, are needed as it gives people born with her anomaly a chance to talk about things they cannot usually talk about without embarrassment.

(d) Evaluation of interview

She felt after the interview as if a burden had been lifted off her shoulders as there are so few people she could talk to about her type of problem.

C. Anorectal malformation (AM 3)

Description of respondent

This respondent was male, 22, married, from social class V and had had numerous residual problems throughout his life concerning incontinence. His colostomy was closed during his teenage years, but had to be re-opened and was finally closed at the age of eighteen.

(a) Effect of residual problems on life (Effect in the past)

(i) Hospitalization

The respondent was an in-patient four times in hospital - at birth for seven months, two to four months at the age of three, three months at the age of 14 and seven months at the age of 17. He could not remember how many operations he had had, but knew that it was more than four. He did not feel that his hospital experience had had an effect on his life.

(ii) Family

He did not think that his residual problems had had an effect on his family, restricted their activities or affected his relationship with them, except for his father. He had a very poor relationship with his father and his residual problems aggravated this. This has continued and when he and his father argued and his father was drunk, he kicked him in the abdomen as he knew it was the weakest part of his body. He had a very good relationship with his mother.

(iii) School

He attended school until standard five. He left because of financial reasons - he had to get a job to help support his family. Although he had a few absences from school, he does not think his residual problems affected his schoolwork. He did have embarrassing incidents at school when his colostomy bag broke. He

was teased about his bag frequently.

(iv) Friends

He had about 13 friends when he was a child and about 13 when he was a teenager - all belonging to a gospel group. He was not friendly with children in his neighbourhood because of his residual problems. These children used to tease him and threaten to pull off his bag or puncture it with a knife. He used to get into many physical fights with these children on this issue. He found the gospel group much more accepting of his problems.

(v) Recreation/social life

His residual problems did restrict his play and sporting activities. He could not do high jump, long jump, javelin or shotput at school. He could not go mountain climbing or ice-skating with his friends as he was often tired and had abdominal pain. He could not stay overnight at friends or go away on holiday because of his colostomy bag - it was too much trouble.

(vi) Dating

He did not go out on many dates when he was a teenager, but said that this was not related to his residual problems. He had his first date when he was eighteen and this was a serious relationship. He does not feel his residual problems had any effect on this relationship. He did not have any sexual relationships when he was a teenager - this choice was not related to his residual problems.

(v) Mental health

He was very depressed about having his illness and used to wonder why he had to be the one to be born with this illness. He used

to feel depressed and anxious often while growing up and smoked cigarettes very often and dagga fairly often. He was dissatisfied with his appearance when he was growing up - because of his colostomy bag. He had no sleeping difficulties or weight problems. The aspect that upset him the most about his residual problems was the way he was teased by other children.

(b) Effect of residual problems on life (Effect at present)

(i) Post-school education and occupational information

He works part-time as a jailkeeper in a magistrate's court. He has been working since the age of twelve in various jobs (mechanic's assistant, labourer, sales assistant and packer). He has had thirteen different jobs. He was unemployed twice, once for a year and once for two months. His residual problems have affected his work in that he cannot do heavy manual work and this is often the only type of work available to him as he only has standard five. The reason he has left many jobs is that he has not had a good relationship with his workmates. He has not experienced any embarrassing incidents at work. He feels that his residual problems have affected his choice of occupation in that he wanted to be a driver, but could not as it involves driving for long periods of time and he cannot be away from a toilet for long.

(ii) Peer relationships

He has over ten friends who are in his gospel band and he last saw one of them a month ago, as they do not live near him. He has no difficulty making friends and does not feel his residual problems have affected his friendships. He has not told them about his problems as he is worried that they will turn against him.

(iii) Marital relationship

This is his first marriage. He does not feel that his residual problems are affecting his present marriage. He has told his wife about his problem but did not tell her before they got married. He feels that his marriage is good and that honesty and communication between them is good.

(iv) Sexual relationship

This is his first sexual relationship which he views as good. His residual problems do not affect how sexually attractive he feels. He does have some problems with sexual functioning but does not know if this is related to his anomaly. He has difficulty getting erections and difficulty ejaculating.

(v) Mental health

He feels satisfied with life as a whole, himself in general and his appearance and could not think of a part of his body which he dislikes most. He often feels depressed and occasionally anxious. He does not have trouble falling asleep. He eats less than usual when he is upset. He often uses alcohol and smokes cigarettes, but never uses dagga. He has never served a jail sentence. He occasionally likes mixing with people. He does not think he has any psychological difficulties at present. He does not feel that any present difficulties (eg. depression) are related to his residual problems, but rather to poor socio-economic circumstances and relationship difficulties with other people.

(vii) Recreation/social life

He rates his social life as very good. He spends most of his spare time reading. He feels that his residual problems restrict him in that he cannot play rugby, soccer or do athletics, but

these are not very important to him.

(c) Medical and Social Work Services

(i) Medical Services

He thinks that he received very good medical and personal treatment from the hospital, especially from the stomatherapist. He was first told about his anomaly when he was nine by his mother. He feels he received sufficient information concerning his anomaly. After the age of fourteen he received medical treatment at Groote Schuur Hospital. He found this move very difficult as he found it embarrassing to tell new doctors about his incontinence. Previously his mother used to tell doctors, but she died when he was twelve. After this he did not say very much to the doctors. He feels that there is a need for a special ward for people born with his anomaly. He did not like being in a ward with other people with other illnesses as they did not have colostomies. He was embarrassed about them seeing his colostomy bag. He did not have any further suggestions concerning medical services, but felt that the doctor should do a home visit once or twice a month to see how a patient copes at home with his problems.

(ii) Social Work Services

He received counselling for depression from a psychiatrist when he was fourteen at the Children's Hospital. He has not felt as if he needed counselling. He does feel that people born with his anomaly need counselling, but could not explain the reason for his answer. He had no suggestions for improving the Social Work Services at the Children's Hospital or in the community.

(e) Evaluation of interview

He said he felt relieved after the interview.

CHAPTER 7

DISCUSSION

Prior to the discussion of the findings (in terms of the research hypothesis) this hypothesis is restated.

7.1 HYPOTHESIS

There is a direct relationship between severity of disability/residual problems and psycho-social functioning, ie the greater the severity, the lower the psycho-social functioning. An assumption, related to the hypothesis, was made at the outset of this study that the high anorectal malformation respondents (AM) would represent the severe disability/residual problem group, the Hirschsprung's disease (HD) respondents would represent the moderate disability/residual problem group and the oesophageal atresia (OA) respondents would represent the mild to no problems disability/residual problem group. Therefore the OA group was expected to have the fewest psycho-social problems, the HD was expected to have some psycho-social problems, and the AM group was expected to have many psycho-social problems.

The discussion section follows the following format:

7.2 DISCUSSION OF VARIABLES (in terms of the research hypothesis)

The first part of the discussion examines certain variables in terms of the research hypothesis in order to determine what influence they have on the hypothesis and to test the hypothesis. This section examines the following:

- A. Medical and health data (This is examined prior to the other variables to see whether the anomaly groups did in fact represent the severity categories into which they were placed at the outset of the study).
- B. Medical information and counselling data

- C. Demographic data
- D. Socio-economic data (Relating to the respondent and the respondent's family).
- E. Family problem data
- F. Psycho-social problem data
- G. Conclusion

7.3 DISCUSSION OF ANOMALY GROUPS IN THE CONTEXT OF THE CONCEPTUAL FRAMEWORK

(in terms of the research hypothesis)

The second part of the discussion examines the three anomaly groups, individually and comparatively, in relation to the research hypothesis and in the context of the conceptual framework. This section examines the following:

- A. Hospitalization, separation and surgery
- B. Developmental stages
- C. Stigma, body image and taboos
- D. Coping

7.4. CONCLUSION.

It must be noted that even though this study examines the psycho-social implications in adulthood of being born with a congenital anomaly, aspects of the respondents' past are discussed in order to put the respondents' present psycho-social functioning in perspective.

This discussion compares the findings of the present study with those in the literature where appropriate. As mentioned in the literature review, no studies were found pertaining to psycho-social effects in adulthood of having the particular anomalies under discussion. Literature pertaining to adults born with other anomalies was examined. It must be emphasized that

these studies of other disability groups cannot strictly be compared with the anomaly groups under discussion. However, as there is no pertinent literature and because there are some common trends between disability groups, this literature is used in the discussion for comparative purposes.

7.2 DISCUSSION OF VARIABLES (IN TERMS OF THE RESEARCH HYPOTHESIS)

A. Medical and health data

(a) General health

General health was rated as good by the majority of respondents in the past and at present by all groups. They rated their health as better than or similar to their health in the past, even though many respondents reported residual problems in the past and at present (discussed later). In view of the number of HD and AM respondents with residual problems, this finding is unexpected. It seems that despite these problems, many respondents viewed their overall health as good.

(b) Nutritional status

The current nutritional status in all groups was adequate in terms of height and weight variables, although the short stature of males in the HD group indicates that they might not have been adequately nourished while growing up. The finding for the OA group was slightly unexpected as this group was expected to have short stature due to early nutritional difficulties. Possibly the fact that the nutritional difficulties in the OA group, although more severe than the HD group, (in the child's early years) did not last for as long as the HD group's nutritional problems accounted for this finding. However, the OA sample is

too small to interpret this data any further. The AM group did not appear to experience any nutritional difficulties, except for a few male respondents.

(c) Residual problems

The OA group had fewer and milder residual problems in the past than the HD and AM group. The HD and AM group reported many residual problems in the past. While the AM group seem to have more bowel problems (proportionately) than the HD group, they seem to have a similar number of respondents with urinary problems. The OA group also had fewer and milder problems at present compared to the HD and AM group.

Residual problems are fewer and milder at present in the HD and AM group than in the past. However, there are still a significant number of respondents with residual problems. The AM and HD group have a similar number of respondents with urinary and bowel problems, but the AM group had more respondents with severe bowel problems.

There seems to be a significant number of respondents reporting sexual functioning problems (30% of those having sexual relationships) and fertility problems (43% of those who have attempted to have children). There was a similar number, proportionately, of HD and AM respondents with sexual and fertility problems (the OA group did not have sexual or fertility problems). No statistics for infertility are available in South Africa, but the international incidence is 10%, ie. 10% of all couples experience some problems with fertility (Speroff et al

1978). The figure in the present study is much higher than this, and although the sample under discussion is very small, this appears to be a significant finding. As the respondents tend to be quite young, perhaps more respondents will have these problems when they get older. Infertility is a controversial issue and both partners need to be assessed. Medical and gynaecological reports concerning some of these respondents seem to indicate that most of the infertility problems in the present study were associated with the surgery to correct the anomaly.

Therefore the HD and AM respondents had residual problems in the past and at present which are socially embarrassing and would put them more at risk than OA respondents for poor psycho-social adjustment.

(d) Restrictions placed on respondents' lives (by residual problems)

Most respondents felt they had been restricted in the past by their residual problems. Most HD and OA respondents no longer feel restricted by their residual problems, while the AM group still view their lives as restricted. This concurs with the medical prognosis for OA and HD respondents, ie. that there will be initial problems (for a longer period in the HD group), but that these problems will diminish over time. The AM group also concurs with the medical prognosis for this group, ie. there will be little improvement in medical health and/or residual problems and they will therefore remain restricted by their problems.

Types of restrictions reported in the past for all groups mainly concern embarrassment over symptoms or not being able to take

part in social or sporting events.

Some HD and AM respondents reported finding social situations very difficult as they could only go to places if they knew there would be easy access to a toilet. One HD respondent described how his mother used to check each time whether there would be a toilet before letting him go on a social outing. An AM respondent recalls that he could not go to sports events at other sportsfields. These problems also seem to occur in ulcerative colitis patients. Reif (1973) notes that people with ulcerative colitis carefully map both routes and places according to the accessibility of toilets. Kelly commenting on his personal experience with ulcerative colitis states, "During adolescence and all through my twenties, certain situations were fraught with danger. Those where there were no toilets in easy reach, in a theatre, cinema or at a sportsground, or on long walks." (Kelly 1986:656) Some respondents reported that they could not stay over at friends as they might have an 'accident', which was also reported by Kelly (1986). In fact for many respondents it was easier to stay at home where they could deal with their problems. This was mentioned in the OA case study, where the respondent reported that his family would prefer not to go to restaurants or out to friends in case he had swallowing or choking difficulties during meals.

Similar types of restrictions were reported in the present by HD and AM respondents, eg. not being able to take part in social or sporting events and embarrassment over symptoms. However, as residual problems were milder there were fewer restrictions on these respondents' lives.

It appears that the OA, HD and AM respondents do represent the severity categories into which they were placed at the outset of the study. The OA group can be seen to represent the no to mild disability/residual problem group. The HD group can be seen to represent the moderate disability/residual problem group. The AM group can be seen to represent severe disability/residual problem group. It is important to note that some of the HD respondents had more severe problems than were initially expected and in fact were not that different in severity to the AM group. However, other HD respondents had no, mild or moderate residual problems, so overall their severity is rated as moderate. (Whereas AM respondents all had high anomalies and were therefore all very severe).

B. Medical information and counselling data

(a) Medical information

The OA and AM groups reported receiving sufficient medical information, but the HD group reported receiving insufficient information, regarding their own medical condition.

Medical information refers to information concerning the anomaly and associated residual problems, received by the respondents from the medical profession and/or their parents while they were growing up.

The reasons for the above findings are not clear and the writer speculates on possible reasons. The OA group probably did not need as much information as the other groups as they were

experiencing few residual problems. The AM group probably received a great deal of information as these respondents were expected to experience residual problems. Furthermore they had more frequent contact with the hospital on account of the severity of their symptoms, and had more exposure to medical staff. The HD group was probably expected to have diminishing residual problems and perhaps was not given as much information as the AM group. The HD group was found to be experiencing more residual problems than was expected, especially later on when they had sexual and fertility problems. The HD group does not seem to have received the medical attention that their problems warranted. As these respondents have recently reached adulthood, some problems have only recently begun surfacing, eg. infertility.

The fact that many respondents reported that they had received insufficient medical information and that many respondents gave incorrect responses when asked about which part of their body was operated on, reveals a great deal of ignorance concerning their anomaly, the operation and associated residual problems. Information was needed on an intellectual level, ie. explaining the aetiology and pathology of the anomaly and the surgical procedures involved and on a practical level, ie. how to cope with the residual problems on a day-to-day basis. Respondents also expressed a need for information on the long-term effects of the anomaly and on possible future problems. It seems that the information was mainly needed during adolescence. Orr et al (1984) note that a general problem among adolescents is concerns about the future.

An important role which the present study played was in identifying respondents who are presently suffering from residual problems, assessing them and if necessary, referring them for medical treatment. Many respondents did not seem to realize that they could be helped. This problem was compounded by the gap between paediatric and adult health services. This seemed to be a problem experienced by many respondents. Respondents commented that even though they were still experiencing problems, they had felt that they were 'too old' for a children's hospital and were unknown when they had gone to an adult hospital. Therefore, they had not returned to either in many cases.

Surgeons need to understand and anticipate the way in which the anomaly will affect the patient. De Wet (1984) points out that some authors have noted that treatment results regarded as good by hospital staff have been accepted with reluctance and far less enthusiasm by parents. Sandberg (1976) notes that it is the psychological meaning rather than the severity of illness per se, which affects the child's personality. Surgeons in consultation with social workers can gain an understanding of the combined effects on patients of physical and psycho-social problems, which will be addressed further in Chapter 8.

This study demonstrated that respondents did have a need for information and advice concerning their residual problems and their day-to-day management.

Knowledge concerning the pathology of these anomalies, surgical techniques and post-operative management has improved

considerably and even at this seemingly late stage, residual problems in all groups can to a certain extent be reduced. Some respondents reported that their quality of life has suddenly improved dramatically since they were identified during the research study as having problems and referred for medical treatment. Present surgical techniques and post-operative management for children born with these anomalies are currently far better than the surgical techniques that were available for the respondents in this study. Furthermore, the surgical follow-up service has improved dramatically. It is unlikely that current patients will experience the type or magnitude of residual problems experienced by the respondents.

(b) Counselling

The OA respondents did not think they had needed counselling in the past, some HD respondents felt they had needed counselling and most AM respondents felt that they had needed counselling (concerning problems with their anomaly).

The reasons that counselling was needed by HD and AM respondents included sexual and fertility concerns, problems with depression, problems with socializing and body image problems. The question age at which counselling was needed yielded minimal response, but those who did reply mentioned adolescence and adulthood as the age at which counselling would have been needed. The fact that many respondents reported needing counselling seems to indicate another gap in services. A few respondents received psychological services during their childhood. However, when they became too old for the paediatric service and did not return

for medical services, they did not appear to return for social work services. This demonstrates the links between the medical and social work service and the need for these bonds to be maintained and strengthened. It appears that the gap in social work service was even greater than the gap in medical services. Some respondents went to adult hospitals for medical treatment, but few seemed to have sought psycho-social services. There needs to be an improvement in the social work follow-up service, which will be examined further in the following chapter.

The taboo nature of the respondents problems could be the reason many respondents, specifically AM respondents, expressed a need for counselling. These respondents probably could not share problems with peers in the usual way as their problems were too embarrassing to share. Johnson (1984) notes that the stigmatization of spina bifida was aggravated in families by the taboos against discussing the condition or anyone's feelings about it. As a result no-one discussed the child's inner concerns with him.

In view of the respondents' need for counselling and the findings concerning psycho-social problems it is clear that social workers have a pivotal role to play in counselling AM and HD patients.

C. Demographic data

(a) Sex/gender variables

There were more males in each group than females, the higher number being in the AM group. As discussed previously, the high

anorectal malformation and Hirschsprung's disease groups have a higher incidence of males than females. There were more males born with oesophageal atresia in the time period covered by the study which accounts for the slight preponderance of males. The importance of respondents' sex in the present study is minimal except that males might have been more concerned with occupational aspects of their lives than female respondents, if they were intending to be the main breadwinner in their families. (The main breadwinner while the respondents were growing up was the father in almost all cases - see Appendix 8 Diagram 8).

(b) Age variables

Most OA and HD respondents are in the youngest age groups, ie. 20-26 while AM respondents tend to be slightly older. The reason for this is not clear. The writer speculates that the overall youth of the total sample is because these respondents had had more recent contact with the hospital than older respondents and were therefore more easily traced. Furthermore, AM respondents, having had many severe residual problems, kept up more contact with the hospital and were more easily traced and therefore included in the study. This could account for their being slightly older than the OA and HD respondents.

When examining the effects of disability/residual problems on a patient's life, the older the patient the more accurate an assessment of psycho-social adjustment becomes (Mattsson 1972). Because of their youth some respondents may not yet have married or begun working and adjustment in terms of these variables can be gauged more accurately when examining older patients. This is

important when assessing the findings of the present study. While findings concerning the OA and HD groups are not incorrect, the findings concerning the AM groups who are slightly older probably provide better indicators of how residual problems have affected their lives.

It is useful to examine age in terms of developmental stages and tasks. Adults in the young adult developmental stage are usually starting to accomplish certain tasks and these need to be considered when examining the following variables listed hereunder (discussed further in the discussion concerning conceptual framework p 177).

(c) Marital status variables

The findings revealed that in all groups, more respondents are single than married, with the greater proportion in the AM group.

This variable was not seen as very useful as regards interpretation concerning adjustment as most of the sample tended to be young. Because of the relative youthfulness of the sample, marital status might depend on a number of factors unrelated to disability/residual problems. The one significant finding was that the tendency to be single was higher in the AM group and as this group tended to be older, being single for these respondents could indicate relationship difficulties. A study on hypospadias found that these adults had a lesser capacity for interpersonal relationships than controls (Berg and Berg 1983). For the HD and OA groups marital status was not found to be an important indicator of adjustment, but it could be more important in the AM

group. Furthermore, those with severe bowel problems were mostly unmarried, which could indicate that residual problems affect interpersonal relationships.

(d) Parental status variables

Concerning children, the findings relating to OA and AM respondents are not very significant, especially in view of the small number involved.

The HD respondents indicate an equal distribution between those with and those without children. These findings need to be considered in conjunction with the findings concerning fertility. Just under half of the married respondents did not have children because these respondents were experiencing problems with fertility as a direct effect of their residual problems. However, as the number involved is very small, this variable is unlikely to have a significant effect on the research hypothesis.

(e) Religious attendance variables

Religious attendance was seen as a possible social support for respondents. The HD respondents practised their religion frequently while the OA and AM respondents did not. This corresponds to their parents' religious attendance for the OA and HD respondents (see Appendix 8 Diagram 2). However, the AM group's religious attendance is low, whereas their parents' was high. The reason for this finding is not clear.

D. Socio-economic data

(a) Educational variables

OA respondents had the lowest education while HD and AM were similar, with AM slightly higher. These findings must be examined in conjunction with the respondents' fathers' education. AM respondents have a similar education to their fathers, HD respondents have slightly higher education than their fathers and OA have similar education to their fathers. Therefore it seems that the education of the respondents is largely determined by the respondents' families' education and there is a strong association between the two. It appears that the education of respondents was not negatively affected by the severity of residual problems.

The educational achievement of the most severe group, ie the AM group, does not support findings in the literature. (This is examined further under socio-economic variables). Orr et al (1984)

in a study of adolescents with chronic illness found that they were more likely to be underachievers at school. Pless and Roghman (1971) in a large epidemiological survey studied children aged 9-11 and concluded that the greater the severity of a condition the greater the extent of underachievement at school. These studies are useful in pointing out the expected effect of greater severity on educational achievement.

(b) Occupational variables

Occupational levels for the purposes of this study are associated with social class. Almost all patients are employed with the exception of 2 who were temporarily unemployed. AM respondents

are in occupations requiring more skills and higher qualifications than HD and OA respondents. These findings must be examined in conjunction with the respondents' fathers' occupations (social class). HD and OA respondents had similar occupations to their families, whereas AM respondents had slightly better occupations (social class) than their families. To a large extent it seems that the occupation of the respondent is determined by the families' social class, although AM respondents are in slightly better occupations. Therefore it seems that the occupation of respondents was not negatively affected by the severity of residual problems.

The occupational achievement of the most severe group, ie the AM group, does not support findings in the literature. Berg et al (1981) found hypospadias to be in occupations requiring lower qualifications than controls.

Although HD and AM respondents seems to have normal occupational achievement, it is not certain whether these respondents' choice of occupation might have been determined by their residual problems. The HD and AM case studies revealed that these respondents were both restricted in their choice of occupation by their residual problems and would have preferred to be in other occupations than the ones they presently hold. Dorner (1977) in a study of spina bifida notes that there is often a gap between ideal work choice and the range of possibilities. Therefore one cannot judge occupational adjustment only in terms of achievement, but also in terms of how the respondent was restricted in his/her choice of occupation.

(c) Income variables

AM have the highest incomes and HD and OA respondents have lower incomes (the latter groups' incomes are similar to each other). Respondents' incomes correspond to their occupation and it was found that severity of residual problems does not appear to have had a negative effect on income levels.

In examining the overall socio-economic status of respondents, it must be noted that social class (ie. occupational level) has been shown to be moderately correlated with education and directly related to qualifications and training (Abrahamson et al 1982). For this reason the three variables are discussed together as a measure of socio-economic status, and will be expanded upon below.

(d) Socio-economic status

It appears that AM respondents have a higher socio-economic status than HD and OA respondents and that all respondents seem to have socio-economic status in keeping with the general population from which they are drawn. In the light of the above findings, the following important facts emerge. Firstly, respondents with severe residual problems (ie. AM) have adjusted well in terms of important life tasks - educational, occupational and financial achievement. In a study of spina bifida adults, Castree and Walker (1981) include independence, occupation and opportunity for social contact with peers in their discussion of the characteristics of an acceptable quality of life. Secondly, these findings are important when the research hypothesis is examined, as socio-economic variables can be eliminated as causal variables of any other psycho-social problems which respondents

experience. Thirdly, the writer speculates as to the reason for the AM group having a higher socio-economic status in the face of debilitating residual problems. Perhaps these respondents, having a handicap, tried to compensate by doing well in their education and occupations. If these respondents had interpersonal relationship problems (reflected in the number of unmarried respondents) and as many of them were socially restricted by their disability (reflected in the discussion of residual problems), achieving in the socio-economic sphere of their lives might have taken on a new importance. De Wet (1984) notes that restrictions placed on extra-mural activities of HD and AM respondents by their residual problems resulted in a one-sided school career consisting only of the academic component.

E. Family problem data

Overall, most respondents did not experience social problems in their families (eg. divorce, substance abuse) while they were growing up. The OA group had no specific problems. The HD group had a few respondents with problem backgrounds and the AM group had the most respondents with problem backgrounds (approximately one-third).

The following questions are addressed: Do severe residual problems cause social problems in a family? Do social problems in a family cause more severe residual problems? These issues are probably simultaneously and reciprocally related.

There seems to be controversy over the role of family factors as a determinant of psycho-social functioning. Harvey and Greenway (1982)

mention that some authors suggest that the effect of poor health and unfavourable family circumstances is cumulative over time, and hence influence behaviour. Heller et al (1985) found that family functioning was not an important determinant of psychological adjustment in children with birth defects. De Wet (1984) found that 50% of the marriages of parents of AM children and 43% of the marriages of parents of HD children were strained. She found little strain (17%) in the marriages of parents of OA children.

A family problem background would be an important issue when examining psycho-social problems experienced by respondents. If respondents exhibited psycho-social maladjustment, this could possibly be due to a family problem background and not necessarily the severity of the residual problems. For example, the AM case study reported being depressed because of social problems in his home (eg. his father's drinking problem), and not because of his residual problems. However, since more respondents in the AM group did not have family problem backgrounds, this variable is probably more useful if viewed as a minor contributory factor to some of the present psycho-social problems experienced by the AM group, not a major determinant. This variable was not found to be very significant in examining the research hypothesis for the OA and HD group.

F. Psycho-social problem data (ie. current problems reported by respondents)

In general most respondents did not have significant problems on various psycho-social dimensions, except for self-esteem and depression. However, there are differences between groups and these are examined. It must also be noted that this data is different to the

demographic, socio-economic and social problem data in that it reports on respondents' subjective perceptions of various psychological and social factors in their lives, whereas the former information was more objective and factual. Psycho-social problem data was obtained by using the Clinical Measurement Package (Hudson 1982). (This was discussed in Chapter 5)

(a) Family relationship problems

Most respondents do not have problems with family relationships. However, just under half the AM respondents do have problems. Therefore in the OA and HD groups, residual problems do not seem to have affected the relationships between respondents and their families, but this might have occurred in the families of some AM respondents.

The finding concerning AM respondents is not that unexpected in that it is substantiated by the data concerning family problems (see p 169). Orr et al (1984) note that family relationships are reported to be worse for youth with chronic illness compared to controls.

However, as discussed previously, Heller et al (1985) did not find family factors to be an important determinant of psychological adjustment. Sigal et al (1973) suggests that severe childhood illness tends to cause later disturbances in the parent-child relationship. This did not seem to occur in the HD and OA group, but did occur in the AM group. This concurs with findings by Gibson (1965) and De Wet (1984), who point out the possible negative effects of anal dilatations in affecting the

parent-child relationship in the AM group.

It would seem that in the AM group, the disruption to the family in terms of socially embarrassing symptoms would be more likely to have a negative effect on relationships between the family and the respondent. This would explain the fact that this group has more problems with family relationships.

(b) Peer relationship problems

There is a tendency for all groups to have no problems with peer relationships, although some AM respondents have problems.

The above findings are expected. The AM respondents probably have more problems with peers than the other groups because of the socially embarrassing nature of their residual problems. The AM case study respondent reported that his residual problems had the most effect on his peer relationships. He was constantly teased about his colostomy when he was a child. Some respondents may also have withdrawn from social events over the years, which was noted by one AM respondent. By avoiding some social situations he had managed to hide his problem from his friends. The fact that some AM respondents can hide their problems probably accounts for the fact that there are only a few respondents in the problem group. It seems that some AM respondents can have fairly normal peer relationships.

The findings concerning HD respondents is slightly unexpected if one considers the residual problems some of them experience. It seems that many HD respondents managed to hide their residual

problems. Furthermore, the most severe residual problems that they were experiencing were sexual and fertility problems which would not be an issue in peer relationships. Overall most respondents in all groups do not have peer relationship problems. This is substantiated in the literature by Orr et al (1984), who found no significant difference in peer relationships in a study of adolescents with chronic illness compared to controls. In a study of spina bifida adults, Laurence and Beresford (1975) noted that incontinence does not affect peer relationships, rather marital relationships.

(c) Self-esteem problems

The majority of AM respondents and HD respondents had problems with self-esteem, while OA respondents did not have problems. As other factors, (eg. poor socio-economic status and family problems) have to some extent been eliminated as major causal factors, it seems that the severity of residual problems has affected self-esteem. The more severe the residual problem, the lower the self-esteem. The least severe group (OA) had the fewest respondents with self-esteem problems, the moderate group (HD) had a significant number of respondents with self-esteem problems and the most severe group (AM) had a majority of respondents with self-esteem problems. It seems likely that the socially embarrassing nature of the residual problems experienced by HD and more so by AM respondents, led to a negative self-concept and lowered self-esteem.

The literature supports the above findings. Adults with hypospadias (Berg and Berg 1983) and adolescents with

myelodysplasia (Hayden et al 1979) were found to have lower self-esteem than controls. Harris (1982) also comments that patients who are self-conscious about abnormalities lack self confidence.

Low self-esteem is experienced because of the stigma and social embarrassment associated with bowel problems (stigma is discussed further on p 180). Cywes (1975) notes that a child's handicap becomes more socially unacceptable as he/she gets older. Teplin et al (1981) note that self-esteem becomes more of an issue to a handicapped child when he/she gets older. Thus adults who are still having bowel problems would be at risk for low self-esteem. Therefore assessing adults' self-esteem is probably a fairly accurate indicator of the effect of residual problems on this psycho-social dimension.

(d) Depression problems

The majority of AM respondents and many HD respondents have problems with depression, while for OA respondents this is not a problem.

Depression is closely associated with self-esteem. It seems that respondents with severe residual problems are more likely to experience depression as other factors (eg. socio-economic and family background) have been largely eliminated as causes of depression.

A significant number of the moderate group (HD) respondents and

the majority of the severe group (AM) respondents reported problems with depression. These findings correspond with self-esteem findings. The socially embarrassing nature of the residual problems and the other associated problems (eg. sexual and/or fertility problems), as well as relationship difficulties and social restrictions might have lowered self-esteem and accounted for these respondents feeling more depressed.

(e) Marital satisfaction problems

There are no significant findings in the AM and OA group as the sub-samples of married respondents in these two groups are very small. Although more HD respondents do not have problems with marital relationships, one-third do have problems.

It is possible that the marital difficulties are associated with sexual or fertility problems and as just under half of the sample of married HD respondents are having problems with fertility, this could be a possible reason for the lack of marital satisfaction. This is substantiated by the HD case-study who saw infertility as being a large problem in her marital relationship. Also, one of the HD respondents said that he would never have got married, had he known that he could not give his wife children.

Laurence and Beresford (1975) note that incontinence does affect marital relationships. In the present study most of the respondents with severe bowel problems were not married - an indicator, perhaps, of the extent to which their bowel problems were affecting their interpersonal relationships.

(f) Sexual satisfaction problems

Concerning sexual satisfaction, few problems were reported in all groups. These findings need to be considered in the context of the findings concerning sexual functioning problems (see p 155). Sexual satisfaction concerns general feelings regarding sexual relationships, rather than the specific problems of sexual function.

One AM respondent reported having sexual satisfaction problems which substantiated the findings regarding sexual problems, ie. 2 respondents reported sexual functioning problems. The HD group seemed to be slightly under-reporting sexual satisfaction problems if one takes into account the number of sexual problems that were reported in the medical questionnaire. The reason for this discrepancy is not clear. Perhaps sexual problems did not interfere with sexual satisfaction, although this seems unlikely. Another possible reason is that the sexual satisfaction questionnaire was administered in the early stages of the research study and respondents might have felt too embarrassed to report difficulties whereas by the time the medical questionnaire was administered, more contact had been established between the writer and the respondent. Possibly respondents, while admitting to other embarrassing problems, ie bowel problems on the medical questionnaire, were not as embarrassed to admit to sexual problems on the medical questionnaire as when they responded to the sexual satisfaction scale (on the psycho-social questionnaire).

7.3 DISCUSSION OF ANOMALY GROUPS IN THE CONTEXT OF THE CONCEPTUAL FRAMEWORK

(and in terms of the research hypothesis)

A. Hospitalization, separation and surgery

It is not clear what effect hospitalization, separation and surgery had on the patients in the study. While the OA group initially had long hospitalization, separation from parents and feeding difficulties, the AM and HD groups probably had more frequent admissions up to a later age than the OA group. In examining the 3 case studies, the hospitalization experience could not be remembered by the OA respondent and in his opinion had a minimal effect on his life. The HD respondent thought the hospitalization experience had a profound, negative effect on her life in terms of separations from the family, friends and absences from school. The AM respondent did not feel that hospitalization had a negative effect on his life. Perhaps this was because he had poor home circumstances and he might have received better physical and emotional care in hospital than at home.

No obvious negative effect of hospitalization, separation and surgery on any of the anomaly groups seems apparent. It is difficult to isolate the effects of hospitalization from the effects of residual problems. This is substantiated in the literature by Gibson's (1965) study on oesophageal atresia and imperforate anus. This study showed no relationship between trauma in early infancy (eg. surgery) and personality adjustment. He notes that the children with the most disruption of feeding and longest separation, ie. the OA group showed the fewest signs of disturbance, as did their mothers. The AM group who had problems relating to defaecation and needed post-surgical anal dilatations, but whose separation was less than the OA group, showed many signs of disturbance, as did their mothers, which Gibson (1965).

and De Wet (1984) relate to the effect of anal dilatations.

While earlier literature (Dowling 1977, Engel and Reichman 1979) indicates that children with feeding problems may have psycho-social problems and disturbed parent-child relationships, a recent study by Lindahl (1984) found OA respondents to have a good psycho-social adjustment to their conditions which was supported by the present study. Referring to surgery the OA group would appear not to have been affected by surgery, whereas the HD and AM group might have been. Blotcky and Grossman's study (1978) found that genito-urinary surgery caused emotional disturbance, but not ENT surgery. The writer speculates that the part of the body operated on in oesophageal atresia

may not be as traumatic as having surgery near the genital region as experienced by the AM group.

B. Developmental stages

Developmental stages and the age of the child were seen as an important aspect in this study.

(a) Early childhood (prior to school)

In terms of Freud's oral developmental stage, where feeding and separation are issues, it appears that in the present study, problems experienced by the OA group at the oral stage of development did not affect respondents' psycho-social functioning negatively.

For the HD and AM respondents, toilet-training difficulties at the anal stage of development might have in some ways been a factor contributing to present psycho-social difficulties. It is

not possible to determine the quantity and quality of this effect.

(b) Childhood

Harris (1981) notes that being self-conscious about an abnormality is usually induced by a specific experience often at a time when self-awareness is important. For example, the HD respondent whose nappy came loose at a scout camp, never returned to scouts. Teplin et al (1981) note that cerebral palsied children seem to retain positive self-esteem similar to non-handicapped children until they reach school. They point out that at this time, the child begins comparing himself/herself to others and this is the time when differences may begin to have a negative effect on self-esteem.

(c) Adolescence

Various authors have noted that as the child gets older, handicap becomes more socially unacceptable (Cywes 1975, Teplin et al 1981, Ditesheim and Templeton 1987). An incontinent adolescent is more socially unacceptable than an incontinent child. Respondents who still had problems at this age found this socially embarrassing.

(d) Adulthood

In terms of achieving some of the life tasks associated with this age as described by Gerdes et al (1981), it is difficult to judge whether the anomaly groups are accomplishing these tasks as they are fairly young. However, the older AM group, while accomplishing occupational tasks adequately, do not seem to be coping well with interpersonal relationship tasks. Some HD

respondents are not coping well with life tasks associated with having children because of fertility problems.

C. Stigma, body-image and taboos

It seems that many of the negative psycho-social effects (of residual problems), ie. low self-esteem and depression, are closely linked with concepts relating to stigma. Socially embarrassing residual problems have caused some HD and AM respondents to feel socially ostracized. This has lowered self-esteem considerably and made them very unhappy. For example one HD respondent tried to commit suicide as his quality of life was so poor because of ongoing problems with incontinence.

The fact that so many respondents tried to hide their problem demonstrates how socially unacceptable these problems were. One HD respondent's wife did not know about his problem. One AM respondent's friends did not know about his problems. The HD case study respondent explained that these issues are "not the sort of thing you want to discuss with others". It is the writer's impression that those respondents who still had socially embarrassing residual problems had a double burden. Firstly, they had to deal with their problem and secondly, they could not discuss their problem easily with others because of its embarrassing nature. The impression the writer gained was that many respondents had for years been burdened by these worries. This was demonstrated in the AM and HD case studies, who felt relieved about being able to talk with someone about their concerns, without feeling embarrassed. Heller et al (1985) note that children with birth defects who are experiencing embarrassment associated with their condition, appear to be the most at risk for psycho-social maladjustment.

Harris (1982) in a study of abnormal appearance, eg. hypertrophic breasts also comments on the distress children felt when they had to go to school, where they had to take part in school activities which they found embarrassing such as organized sport and physical education. This was also an issue for respondents in the present study, not only because some of them had colostomies but also, as explicitly described in the case study, because of scarring. Although colostomies and scarring could usually be hidden, when changing into gym clothes they were not always successfully hidden.

The HD case study and one of the older HD respondents in his medical questionnaire commented on the negative effect their scars had had for them and problems relating to their body image. The writer questions whether scarring was a problem for other HD and AM respondents. As this was an exploratory study only some aspects regarding residual problems were examined. These problem areas mainly concerned bowel and sexual/fertility problems. The HD case study was important in showing that scarring might have been as distressing and socially embarrassing as other residual problems.

In some ways the secrecy and initially unnoticeable nature of residual problems experienced by HD and AM respondents, seem to produce conflicting feelings. On the one hand, respondents reported trying to cover up their problems. Kelly (1986) and Reif (1973) in commenting on ulcerative colitis note the many methods used by patients to cover up odour and excrement. To a certain extent, their disability was unseen, but if respondents had an accident, it became very obvious. It seems respondents could have a fairly normal life until they were 'found out'. For example, one HD respondent had a girlfriend, but she

left him when she discovered his problem. On the other hand, some respondents wished their disability were more obvious. The HD case study respondent explained that as her bowel problems (eg. constipation), were largely unseen, people expected certain behaviour from her, eg. to participate in sport, as would a person without physical problems. Sometimes she wished her disability were more obvious. Tavormina et al (1976) describe a similar situation in discussing diabetics whom, they explain, have no visible physical disability and therefore find difficulty in convincing their peers that they cannot keep up with physical activities.

D. Coping

Although the AM and HD respondents experienced socially embarrassing problems they seemed to have worked out various strategies and defences to deal with these concerns (see Appendix 9 for a description of practical coping strategies). However, coping was not really examined in any detail in the present study and will not be discussed further.

7.4 CONCLUSION

The present study supported the research hypothesis to a large extent and showed that more severe disability/residual problems had a negative effect on psycho-social functioning. It seemed to effect psychological functioning, but had little effect on socio-economic functioning.

Most of the AM respondents (most severe), were found to be having psychological problems, ie. low self-esteem and depression, some interpersonal problems and some problems concerning restrictions of their social lives. Some of the HD respondents (moderate) were found to be having psychological problems, ie. low self-esteem and depression,

interpersonal relationship difficulties and a restricted social life. The OA group (no to mild problems) were having no psycho-social problems.

While experiencing other psycho-social problems, AM respondents have coped better in this aspect of their lives than OA and HD respondents. However, it must be noted that socio-economic status in all groups seemed to correspond with respondents' families' socio-economic status. It appeared that residual problems did not effect this aspect of respondents' lives.

It therefore appears that severity of residual problems in the AM and HD groups had the most effect on self-esteem, depression and interpersonal relationships and the least effect on peer relationships, family relationships and sexual satisfaction. Residual problems seemed to have had a minimal or non-existent effect on the lives of OA respondents.

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The fact that severity of disability causes poorer psycho-social adjustment is supported by many studies in the literature (Pless and Roghman 1971, Stein and Jessop 1984, Sandberg 1976, Heller et al 1971). However, there are studies which cite other factors such as parental adaptation (Kolin et al 1971) developmental stages and quality of parent-child relationships (Mattsson 1972) as more important determinants of psycho-social adjustment than severity of illness/disability. However, it must be noted that all these studies mainly concern children and adolescents. Probably as respondents with embarrassing residual problems get older, family factors lose their former significance and severity of residual problems with their accompanying sequelae of lower self-esteem, depression and interpersonal difficulties become more important in determining psycho-social functioning.

Concerning medical and counselling data, the findings reveal gaps in the medical and social work services. It seems that some respondents did not receive sufficient medical information concerning their anomaly and did not receive an adequate follow-up service. In regard to social work services, there appeared to be a need by some respondents for counselling and a need for an improved follow-up service.

Conclusions and recommendations pertaining to the dissertation, appear in the following chapter.

CHAPTER 8CONCLUSIONS AND RECOMMENDATIONS8.1 CONCLUSIONS

The following chapter presents the conclusions reached in the study and recommendations based on these conclusions.

Where surgery for the congenital anomalies under discussion leaves patients with residual problems, the associated psycho-social sequelae frequently persist into adulthood.

The literature concerning the psycho-social effects of being born with these anomalies indicate that patients born with Hirschsprung's disease and anorectal malformations are at risk for psycho-social maladjustment. This risk appears minimal for patients born with oesophageal atresia.

The writer found that the three anomaly groups appear to have the psycho-social functioning that was expected from them in terms of their medical prognosis and the literature. Despite early feeding difficulties, hospitalization and some residual problems, the psycho-social functioning of oesophageal atresia respondents did not seem to be affected to any great extent. However, due to residual problems and other possible factors, eg. more frequent hospital admissions, toilet training difficulties, many anorectal malformation and some Hirschsprung's disease respondents can be seen to be at risk for psycho-social maladjustment.

There is a lack of consensus amongst studies concerning the psycho-social effects of severity of disability/illness. The present study supported the

research hypothesis to a large extent and showed that the more severe disability/residual problems had a negative effect on psycho-social functioning.

This concurs with many studies in the literature. The studies that cite other factors, eg. parent/child relationships as being more important determinants of psycho-social functioning, mainly concern children and adolescents. However, as other authors indicate, disability becomes more socially unacceptable as adulthood approaches. This could explain why parent/child relationships lose their former importance and the severity of disability/illness becomes more significant. This is especially so if residual problems are stigmatizing, as are the socially embarrassing symptoms associated with bowel disorders as found in the study.

More specifically, severe residual problems had a negative effect on psycho-social functioning, ie. self-esteem and depression and interpersonal variables and social functioning, ie. social/recreational aspects, as demonstrated in the AM and to a lesser extent the HD group. Severity seems to have had less of an effect on peer relationships, family relationships and sexual satisfaction. Severity of disability/residual problems seems to have had little effect on socio-economic factors which appear to be related to families' socio-economic status. Residual problems seem to have had a minimal or non-existent effect on the psycho-social functioning of the OA group.

The study reveals that patients do not always receive the medical and social work services that are required for those dealing with severe residual problems. Although implications for medical and social work are largely discussed separately, some of the recommendations for the two are

discussed in conjunction with each other - demonstrating the importance of links between them. The importance of a multi-disciplinary team approach to health care of patients with disability/illness has been repeatedly emphasized in the literature (Kanthor et al 1974, Bax et al 1988, Feldman 1974).

8.2 IMPLICATIONS AND RECOMMENDATIONS FOR THE MEDICAL AND SOCIAL WORK PROFESSIONS

A. Medical services

As many patients have residual problems and lacked medical information regarding their condition and its day-to-day management, the medical profession have important tasks to perform. An examination of the resources and facilities available for these patients and the findings of the present study regarding follow-up services, indicate gaps in medical services. There especially needs to be an improvement in services for the HD and AM patients.

While this situation had improved recently, ongoing attention must be paid to this vital aspect of patient care. Follow-up could help to eliminate many residual problems and therefore many associated psycho-social problems.

As there are at present no specialized adolescent services available for these patients, other strategies need to be employed to bridge the gap between paediatric and adult health care. Close liaison between surgeons at the paediatric and adult hospital, especially during the initial transition period is important. This might alleviate the anonymity of large adult hospitals. It is recommended that a paediatric surgeon and the surgeon at the adult hospital initially see the patient together during this transition period. Handing over this

responsibility to the adult hospital would lessen the present burden for the paediatric service (which is presently following up these patients).

Specifically, more medical attention should be paid to residual problems in the HD group as some respondents have more severe residual problems than their medical prognosis would indicate. Here social workers could play a role in determining whether physical problems were due to organic or psycho-social factors and could advise surgeons in this regard.

Regarding medical information received, it appears that respondents were given information in accordance with their medical prognosis. The HD group was not expected to have severe residual problems, hence the information revealed by some respondents was reported to be insufficient.

Despite the fact that many respondents felt that they had received sufficient information, further questioning revealed ignorance regarding their condition and medical treatment. Practical advice and theoretical information about the aetiology and long-term effects of disability/residual problems, was needed by all groups. A need for this type of information was also reported in other studies. It seems that information is needed at all ages, but especially during adolescence. Part of the doctors' role should be to pass on information to their patients and this does not seem to be taking place sufficiently. Where doctors do attempt this educative role, they generally inform the parents of the patient about the condition. It is most important that the patient should be given the information

personally. At each visit, the doctor should check that both the patient and the parents understand the medical information imparted.

Surgeons should have a specific programme providing medical information relevant to the patients' special needs and problems at various ages. Information should be given before a patient starts dealing with certain issues, eg. information concerning possible sexual and/or fertility problems should be given before the patient begins sexual relationships and before he/she starts attempting to have children. While information could be standardized to a certain extent, differences in severity and types of problems experienced, would mean that each individual's special needs regarding information should be considered. Information regarding the aetiology and pathology of the anomaly, the operative procedures and the long-term effects must be given. While adolescence was mentioned as an age when information was needed, the writer proposes that information be given to respondents on an ongoing basis and at a level appropriate to the patient's developmental age, eg. use of illustrations for young children. The social worker could provide information regarding cognitive levels and together with surgeons decide which information would be appropriate for patients.

The risks, however small, associated with later physical problems related to sexuality and fertility need to be pointed out to patients during adolescence. As this is an emotion-laden topic perhaps social workers could be involved in this process.

B. Social work services

There appears to be a great need for social work services; many HD and AM respondents had psycho-social problems and expressed a need for counselling. Few respondents had received counselling. In terms of the taboo nature of the problems experienced by these respondents, social workers have a crucial role to play in counselling. OA respondents, however, did not appear to need social work services. Even though current residual problems are probably milder due to improved medical services, the HD and AM patients could still be at risk for psycho-social adjustment and might need social work services.

While the paediatric surgery service for these patients has improved dramatically, improvement in social work services has not been as impressive. Attention needs to be given to social work resources and facilities for these patients. Follow-up services are especially lacking. Bracht (1978) quotes Clarke: 'Scientific strides in the biological, physiological, chemical and pharmaceutical components in preventing and treating disease have not been matched with corresponding advances in psycho-social treatment and prevention of dysfunction. The ability to help with the psychological, sociological and physical consequences of illness has not kept pace with the technological ability to sustain and prolong life' (Bracht 1978: 46).

Bartlett (1961) views the social worker's role as encompassing 2 orientations, firstly toward the patient and his/her family and secondly toward the medical team.

Regarding the former, Bartlett (1961) points out that while other workers in the health field are concerned with psycho-social factors,

the social worker is the only person with a consistent focus on social functioning. It would be the social worker's responsibility to identify, through consultation with the medical staff, those patients at risk for psycho-social maladjustment.

Social workers should concentrate on dealing with self-esteem, depression and interpersonal difficulties. They might need to work through feelings associated with their self-concept and body-image, to build up their self-confidence. It is recommended that this individual counselling be combined with group therapy.

Self-help groups would be a feasible support for these patients. A group in which patients could talk about embarrassing problems without feeling ostracized may be a very useful therapeutic method. It would be useful if this group were not only aimed at therapeutic goals, but also educative ones. They could share information regarding how to cope with certain problems and difficult situations. Medical personnel could attend some group meetings to provide information, eg. stomatherapists, medical representatives who deal with incontinence aids.

These groups might provide an important support system in view of the taboo nature of these respondents' problems. This applies especially to those patients who could not talk about their concerns to anyone else. For patients who had been previously unable to share feelings and experiences, this catharsis might prove to be very beneficial. In this group, patients could feel 'safe' to talk about intimate topics without embarrassment.

The writer proposes that a useful team approach to counselling for HD and AM patients would be in the form of a social worker/stomatherapist team. Many of the respondents in the study expressed concern over managing embarrassing residual problems. The stomatherapist could provide factual medical information, eg. dietary advice and practical strategies for dealing with symptoms. The social worker could help 'slot' these strategies into various aspects of the patient's life. For example, the social worker and patient might decide on how the patient could cope with certain social situations, based on strategies recommended by the stomatherapist. This might mean that patients are able to take part in various social or sporting activities which they previously avoided, because they were unable to manage their symptoms. If some restrictions on patients' lives were eliminated, this could considerably improve patients' quality of life. This could lead to fewer psycho-social difficulties.

Respondents are coping well with socio-economic aspects in their lives. However, should they be experiencing problems, eg. in their occupations, the social worker/stomatherapist team could provide some practical advice on how to alleviate difficulties. They could also ensure that respondents are placed in occupations suited to their special needs. If necessary, they might need to educate patients' work colleagues about these patients' special needs and problems. This could lessen social ostracism in the work place, but would need to be handled with sensitivity so as not to aggravate the situation.

Those who had other psycho-social concerns, not directly related to residual problems, could receive individual counselling and/or group therapy.

In conclusion, it is obvious from this study that physical residual problems and psycho-social functioning are intertwined. As the hypothesis was supported, a reduction in residual problems, ie. less severity should lead to fewer psycho-social problems. Team-work between medical and social work professions is therefore essential. Social workers and surgeons must act in a preventive capacity when they have identified risk situations to minimize the negative effects on psycho-social functioning.

Since the paediatric surgery service has improved considerably, patients currently being operated upon for these anomalies should have fewer residual and probably fewer psycho-social problems than the respondents in the present study. Under these circumstances, the size and scope of the social work service for these patients would be reduced.

8.3 RECOMMENDATIONS FOR ADDITIONAL RESEARCH

This study has revealed some areas which require further examination:

- A. Coping was only examined briefly in the present study. Research on the practical techniques used to manage symptoms and the psycho-social strategies used to manage patients' lives would be worthwhile. Regarding the latter, this research could specifically focus on the support systems used by patients.
- B. The present study suggested that scarring might have as negative an effect on psycho-social functioning as bowel, sexual or fertility problems. It would be important to investigate the effects of scarring on the body-image and self-esteem of patients.

- C. The present study examined the psycho-social implications in adulthood of being born with a congenital anomaly. A previous study (De Wet 1984), examined the psycho-social effect of congenital anomalies on the family and to a certain extent on the child. The psycho-social effects of these anomalies in adolescence have not been examined. By adolescence, OA patients are generally free of residual problems, whilst HD patients are learning to manage their problems. AM patients, however, have persisting residual problems. In view of the above it would be useful to explore any physical and psycho-social problems at this age.
- D. Related to the above issue, the feasibility of an adolescent unit for patients with chronic diseases should be explored to bridge the gap between paediatric and adult health care.

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APPENDIX 1

GLOSSARY

Aganglionosis	-	Congenital absence of ganglion (nerve) cells.
Anastomotic	-	The surgical joining of two hollow ends after creating an opening.
Aspiration	-	Removal of fluids or gases from a cavity.
Biopsy	-	Removal of tissue or other material from the body for diagnostic purposes.
Colostomy	-	The surgical creation of a new opening of the colon on the surface of the body.
Dilatation	-	The act of stretching or expanding a cavity.
Distal	-	Remote, furthest from a point.
Enterocolitis	-	Inflammation of the small intestine and colon.
Epiphyses	-	The wide growth ends of the long bones.
Fistula	-	An abnormal passage usually between two internal organs, but also from an internal organ to the body's surface.
Ganglion	-	A group of nerve cell bodies.
Gastrostomy	-	A surgical creation of an opening into the stomach.
Ileostomy	-	A surgical creation of a new opening of the ileum on the surface of the body.
Imperforate	-	Abnormally closed.
Meconium	-	The dark-green contents of the intestine of the full-term fetus.
Necrotizing	-	Causing death of tissue.
Perineum	-	Space between anus and scrotum.
Proximal	-	Nearest, closest to a point.
Resection	-	Excision of a portion of an organ or structure.
Stenosis	-	Narrowing of a canal.
Urethra	-	The canal which conveys urine from the bladder to the exterior of the body.
Vestibular	-	Pertaining to a space or cavity at the entrance to a canal.

APPENDIX 2

QUESTIONNAIRE A

ADULT STUDY (Red Cross War Memorial Children's Hospital)

We are very interested in knowing how our past patients, who had operations at this hospital, are doing. We would be very grateful if you could help us with this information. This information is confidential and your name will not be used under any circumstances.

INSTRUCTIONS

Please fill in all the questions with one of the following:

- a tick ()
- a short answer
- a number

depending on what is required.

Thank you for your help.

Study No.

--	--

PRESENT CIRCUMSTANCES

1. Date of birth (day/month/year)

--	--	--	--	--	--

2. How old are you, in years?

--	--

3. Are you male or female ? (Tick the correct box)

male	
female	

4. What is your marital status ?

married	
living together	
divorced	
separated	
widowed	
single	

5. How many children do you have ? (Include stepchildren)

none	
one	
two	
three	
more than three	

6. To what religion do you belong ?

7. Approximately how often do you attend religious services ?

more than twice per week	
twice per week	
two to three times per month	
once per month	
once every few months	
never	

8. What is your home language ?

English	
Afrikaans	
English and Afrikaans	
other	

9. At present, who are you living with? (i.e. who lives with you in your household eg. parents, friends, wife, etc.)

.....

10. In which suburb do you live ?

11. What type of place do you live in ?

self-owned house	
self-owned flat	
rented house	
rented flat	
rented room	
other	
no fixed home	

12. How long have you been staying in the above ?
(Indicate whether this is in months/years)

13. Are you staying in a household ?
(with other people eg. parents, friends, wife, etc.)

yes	
no	

(If yes, go to question 14)

(If no, go to question 17)

14. How many rooms are used for sleeping in your
household ?

--	--

15. How many people are there living with you under 10 years
old ?

--	--

16. How many people are there living with you who are 10 years
old and over ?

--	--

17. What is the highest school standard you have passed ?

(If you did not go to school, mark the box with 0)

highest standard passed

--	--

18. Have you any post-school qualifications ?

(If no, go to question 20)

yes	
no	

19. If yes, please describe them:

.....

20. Are you employed at present ?

(If no, go to question 22)

yes	
no	

21. If yes, describe your work:

.....

22. What is your income after deductions ? (indicate whether the amount is per week or per month)

23. What is your household's combined income after deductions ?

(indicate whether the amount is per week or per month)

PAST CIRCUMSTANCES: In the following questions the words 'when you were growing up' means when you were 19 years old and under i.e. 0-19 years old.

24. What was your father's main occupation (work) when you were growing up ?

.....

25. What was your mother's main occupation (work) when you were growing up ?

.....

26. Were your parents employed most of the time when you were growing up?

	mother	father
all of the time		
most of the time		
some of the time		
hardly ever		
never		

27. What is the highest standard at school that your mother and father passed ? (if they did not go to school, indicate with 0)

	mother	father
highest standard passed		

28. Do your parents have any post-school qualifications ?

	mother	father
yes		
no		

29. Who was the main breadwinner in your house when you were growing up ?

.....

30. How well off do you think your family/household was when you were growing up compared to your friends ?

much better off than friends	
better off than friends	
about the same as friends	
less well off than friends	
much less well off than friends	

31. Who did you mostly live with when you were growing up ?

both parents	
one parent	
neither parent, but relative	
neither parent, but friend	
foster parents	
adoptive parents	
childrens home	
other	

32. If your mother and father are divorced or separated, how many years old were you when the first separation occurred ? (place an 0 in the box if your parents were not separated or divorced)

--	--

33. Are both of your parents still living ?

	mother	father
yes		
no		

34. Approximately how many brothers and sisters (including stepbrothers and sisters) stayed in your household when you were growing up ?

brothers	
sisters	

35. How religious would you say your parents were ?

very religious	
moderately religious	
slightly religious	
not religious	

36. Do you think there were social problems in your household when you were growing up ?

(If no, go to question 38)

yes	
no	

37. If yes, describe them:

.....

.....

.....

.....

38. Did your parents ever receive counselling or psychiatric help when you were growing up ?

	mother	father
no		
yes		
don't know		

39. Was your mother or father ever convicted of a criminal offence ?

mother was convicted	
father was convicted	
both were convicted	
neither were convicted	
don't know	

40. In your opinion, how much alcohol does or did your mother and father drink ?

	mother	father
never drinks		
sometimes drinks a little		
drinks in moderation		
drinks quite a lot		
drinks far too much		
don't know		

41. In your opinion, how often does or did your mother and father use drugs?

	mother	father
never uses them		
sometimes uses a little		
uses them often		
uses them too much		
uses them far too much		
don't know		

42. In your opinion, how often did your father/stepfather and mother/step-mother use physical violence on you ?

	mother	father
very often		
often		
occasionally		
rarely		
never		

43. In your opinion, how often did your father/stepfather use physical violence on your mother and brothers and sisters ?

	on mother	on brothers/sisters
very often		
often		
occasionally		
rarely		
never		

44. How often did your family move houses ?

very often	
often	
occasionally	
rarely	
never	

45. Would you generally describe your childhood as a happy one ?

yes	
no	

46. Please explain the reason for your answer above (question 45)

.....

.....

.....

.....

.....

.....

.....

Please check to see you have answered everything.

Thank you again for your help.

CLINICAL MEASUREMENT PACKAGE

Hudson
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QUESTIONNAIRE B

INDEX OF PEER RELATIONS (IPR)

Study No _____

This questionnaire is designed to measure the way you feel about the people you work, play or associate with most of the time; your peer group. It is not a test so there are no right or wrong answers. Answer each item as carefully and as accurately as you can by placing a number beside each one as follows:

1. Rarely or none of the time
2. A little of the time
3. Some of the time
4. A good part of the time
5. Most or all of the time

please begin.

1. I get along very well with my peers
2. My peers act like they don't care about me
3. My peers treat me badly
4. My peers really seem to respect me
5. I don't feel like I am 'part of the group'
6. My peers are a bunch of snobs
7. My peers understand me
8. My peers seem to like me very much
9. I really feel 'left out' of my peer group
10. I hate my present peer group
11. My peers seem to like having me around
12. I really like my present peer group
13. I really feel like I am disliked by my peers
14. I wish I had a different peer group
15. My peers are very nice to me
16. My peers seem to look up to me
17. My peers think I am important to them
18. My peers are a real source of pleasure to me
19. My peers don't seem to even notice me
20. I wish I were not part of this peer group
21. My peers regard my ideas and opinions very highly
22. I feel like I am an important member of my peer group
23. I can't stand to be around my peer group
24. My peers seem to look down on me
25. My peers really do not interest me

[illegible]

,4,7,8,11,12,15,16,17,18,21,22

1. Rarely or none of the time
2. A little of the time
3. Some of the time
4. A good part of the time
5. Most or all of the time

1. I feel that people would not like me if they really knew me well
2. I feel that others get along much better than I do
3. I feel that I am a beautiful person
4. When I am with other people I feel they are glad I am with them
5. I feel that people really like to talk with me
6. I feel that I am a very competent person
7. I think I make a good impression on others
8. I feel that I need more self-confidence
9. When I am with strangers I am very nervous
10. I think that I am a dull person
11. I feel ugly
12. I feel that others have more fun than I do
13. I feel that I bore people
14. I think my friends find me interesting
15. I think I have a good sense of humour
16. I feel very self-conscious when I am with strangers
17. I feel that if I could be more like other people I would have it made
18. I feel that people have a good time when they are with me
19. I feel like a wallflower when I go out
20. I feel I get pushed around more than others
21. I think I am a rather nice person.
22. I feel that people really like me very much
23. I feel that I am a likeable person
24. I am afraid I will appear foolish to others
25. My friends think very highly of me

[illegible]

NOTE: If you are unmarried at present, do not answer this questionnaire

INDEX OF MARITAL SATISFACTION (IMS)

Study No .

This questionnaire is designed to measure the degree of satisfaction you have with your present marriage. It is not a test, so there are no right or wrong answers. Answer each item as carefully and as accurately as you can by placing a number beside each one as follows:

1. Rarely or none of the time
2. A little of the time
3. Some of the time
4. A good part of the time
5. Most or all of the time

Please begin.

1. I feel that my partner is affectionate enough
2. I feel that my partner treats me badly
3. I feel that my partner really cares for me
4. I feel that I would not choose the same partner if I had to do it over again
5. I feel that I can trust my partner
6. I feel that our relationship is breaking up
7. I feel that my partner doesn't understand me
8. I feel that our relationship is a good one
9. I feel that ours is a very happy relationship
10. I feel that our life together is dull
11. I feel that we have a lot of fun together
12. I feel that my partner doesn't confide in me
13. I feel that ours is a very close relationship
14. I feel that I cannot rely on my partner
15. I feel that we do not have enough interests in common
16. I feel that we manage arguments and disagreements very well
17. I feel that we do a good job of managing our finances
18. I feel that I should never have married my partner
19. I feel that my partner and I get along very well together
20. I feel that our relationship is very stable
21. I feel that my partner is a comfort to me
22. I feel that I no longer care for my partner
23. I feel that the future looks bright for our relationship
24. I feel that our relationship is empty
25. I feel there is no excitement in our relationship

[illegible]

1, 3, 5, 8, 9, 11, 13, 16, 17, 19, 20, 21, 23

NOTE: Do not answer this questionnaire if you have never had a sexual relationship. If you are presently having a sexual relationship, answer this questionnaire according to your present sexual partner. If you are not presently having a sexual relationship, but have had one in the past, answer the questions according to your last partner.

INDEX OF SEXUAL SATISFACTION (ISS) Study No _____

This questionnaire is designed to measure the degree of satisfaction you have in the sexual relationship with your partner. It is not a test, so there are no right or wrong answers. Answer each item as carefully and accurately as you can by placing a number beside each one as follows:

- 1. Rarely or none of the time
- 2. A little of the time
- 3. Some of the time
- 4. A good part of the time
- 5. Most or all of the time

Please begin.

- 1. I feel that my partner enjoys our sex life
- 2. My sex life is very exciting
- 3. Sex is fun for my partner and me
- 4. Sex with my partner has become a chore for me
- 5. I feel that sex is dirty and disgusting
- 6. My sex life is monotonous
- 7. When we have sex it is too rushed and hurriedly completed
- 8. I feel that my sex life is lacking in quality
- 9. My partner is sexually very exciting
- 10. I enjoy the sex techniques that my partner likes or uses
- 11. I feel that my partner wants too much sex from me
- 12. I think that sex is wonderful
- 13. My partner dwells on sex too much
- 14. I try to avoid sexual contact with my partner
- 15. My partner is too rough or brutal when we have sex
- 16. My partner is a wonderful sex mate
- 17. I feel that sex is a normal function of our relationship
- 18. My partner does not want sex when I do
- 19. I feel that our sex life really adds a lot to our relationship
- 20. My partner seems to avoid sexual contact with me
- 21. It is easy for me to get sexually excited by my partner
- 22. I feel that my partner is sexually pleased with me
- 23. My partner is very sensitive to my sexual needs and desires
- 24. My partner does not satisfy me sexually
- 25. I feel that my sex life is boring

Study No

1. Rarely or none of the time
2. A little of the time
3. Some of the time
4. A good part of the time
5. Most or all of the time

1. I feel powerless to do anything about my life
2. I feel blue
3. I am restless and can't keep still
4. I have crying spells
5. It is easy for me to relax
6. I have a hard time getting started on things that I need to do
7. I do not sleep well at night
8. When things get tough, I feel there is always someone I can turn to
9. I feel that the future looks bright for me
10. I feel downhearted
11. I feel that I am needed
12. I feel that I am appreciated by others
13. I enjoy being active and busy
14. I feel that others would be better off without me
15. I enjoy being with other people
16. I feel it is easy for me to make decisions
17. I feel downtrodden
18. I am irritable
19. I get upset easily
20. It is hard for me to have a good time
21. I have a full life
22. I feel that people really care about me
23. I have a great deal of fun
24. I feel great in the morning
25. I feel that my situation is hopeless

[illegible]

1. Rarely or none of the time
2. A little of the time
3. Some of the time
4. A good part of the time
5. Most or all of the time

1. The members of my family really care about each other
2. I think my family is terrific
3. My family gets on my nerves
4. I really enjoy my family
5. I can really depend on my family
6. I really do not care to be around my family
7. I wish I was not part of this family
8. I get along well with my family
9. Members of my family argue too much
10. There is no sense of closeness in my family
11. I feel like a stranger in my family
12. My family does not understand me
13. There is too much hatred in my family
14. Members of my family are really good to one another
15. My family is well respected by those who know us
16. There seems to be a lot of friction in my family
17. There is a lot of love in my family
18. Members of my family get along well together
19. Life in my family is generally unpleasant
20. My family is a great joy to me
21. I feel proud of my family
22. Other families seem to get along better than ours
23. My family is a real source of comfort to me
24. I feel left out of my family
25. My family is an unhappy one

[illegible]

1, 2, 4, 5, 8, 14, 15, 17, 18, 20, 21, 23

APPENDIX 4

QUESTIONNAIRE C

ADULT STUDY (Red Cross War Memorial Children's Hospital)

The Department of Paediatric Surgery is very interested in knowing about the health of its past patients who had operations at this hospital. This questionnaire asks about your past health ie. while you were growing up (Section A) and about your present health (Section B). We would be very grateful if you could help us with this information. This information is confidential and your name will not be used under any circumstances.

This questionnaire is also being given to patients who had different illnesses to yours and who had different operations so some of the questions may seem irrelevant to you - nevertheless please answer all questions.

Thank you for your help.

A. INFORMATION CONCERNING YOUR HEALTH IN THE PAST

Study No _____

1. Overall how would you see your health while you were growing up?
(tick the appropriate box)

very poor	
poor	
good	
very good	

2. On what part of your body did you have your operations (at Red Cross Hospital) when you were a baby/child?

.....

3. Did you have any problems with that part of your body while growing up?
(if no, go to question 5)

yes	
no	

4. If yes, describe these problems

.....

.....

.....

.....

5. Did you experience any of the following problems on a regular basis while growing up?
(if they were never a problem, place an 0 next to the problem).

...../ -2-

swallowing difficulties	
choking while eating	
special diet/foods	
abnormal coughing	
chest infections	
no control over your wind	
constipation	
diarrhoea (upset stomach)	
smearing in pants	
no control over bowels	
wind in abdomen/stomach	
no control over bladder	
dribbling urine/water	
burning when passing urine/water	
bladder infections	
problems with sexual functioning	
abdominal/stomach pain	

6. At what age did you find these difficulties most problematic (if they were never a problem place an 0 next to the problem)

Swallowing difficulties
Choking while eating
Special diet/foods
Abnormal coughing
Chest infections
No control over your wind
Constipation
Diarrhoea (upset stomach)
Smearing in pants
No control over bowels
Wind in abdomen/stomach
No control over bladder
Dribbling urine/water
Burning when passing urine/water
Bladder infections
Problems with sexual functioning
Abdominal/stomach pain

7. How often did these difficulties occur? (if they were never a problem place an 0 next to the problem)

Swallowing difficulties
 Choking while eating
 Special diet/foods
 Abnormal coughing
 Chest infections
 No control over your wind
 Constipation
 Diarrhoea (upset stomach)
 Smearing in pants
 No control over bowels
 Wind in abdomen/stomach
 No control over bladder
 Dribbling urine/water
 Burning when passing urine/water
 Bladder infections
 Problems with sexual functioning
 Abdominal/stomach pain

8. At what age did these difficulties stop?
 (if they were never a problem place an 0 next to the problem, if they haven't stopped place an X next to the problem)

Swallowing difficulties
 Choking while eating
 Special diet/foods
 Abnormal coughing
 Chest infections
 No control over your wind
 Constipation
 Diarrhoea (upset stomach)
 Smearing in pants
 No control over bowels
 Wind in abdomen/stomach
 No control over bladder
 Dribbling urine/water
 Burning when passing urine/water
 Bladder infections
 Problems with sexual functioning
 Abdominal/stomach pain

9. How did you handle or cope with these problems, eg. what treatments, medication, and aids did you use? (if they were never a problem place an 0 next to the problem)

Swallowing difficulties
 Choking while eating
 Special diet/foods
 Abnormal coughing
 Chest infections
 No control over your wind
 Constipation
 Diarrhoea (upset stomach)
 Smearing in pants
 No control over bowels
 Wind in abdomen/stomach
 No control over bladder
 Dribbling urine/water
 Burning when passing urine/water
 Bladder infections
 Problems with sexual functioning
 Abdominal/stomach pain

10. Did your physical difficulties restrict your life in any way while you were growing up? eg. socially, at school, sports, on holiday, etc. (if no, go to question B.1)

yes	
no	

11. If yes, describe this

.....

B. INFORMATION CONCERNING PRESENT HEALTH

1. How do you see your health at present?

very poor	
poor	
good	
very good	

2. How much do you weigh?

--	--	--

 kg

or

--	--	--

 pounds/lbs

3. How tall are you? (approximately)

4. Describe any illnesses or physical complaints you have at present

.....

.....

.....

5. Describe any operations you have had since your last operation at Red Cross Hospital

.....

.....

.....

6. Are you still having problems with that part of your body which was operated on when you were a child? (if no, go to question 8)

yes	
no	

7. If yes, describe these problems

.....

.....

.....

.....

8. Do you experience any of the following problems on a regular basis, at present (if they are never a problem place an 0 next to the problem)

Swallowing difficulties	
Choking while eating	
Special diet/foods	
Abnormal coughing	
Chest infections	
No control over your wind	
Constipation	
Diarrhoea (upset stomach)	
Smearing in pants	
No control over bowels	
Wind in abdomen/stomach	
No control over bladder	
Dribbling urine/water	
Burning when passing urine/water	
Bladder infections	
Problems with sexual functioning	
Abdominal/stomach pain	

9. How often do these problems occur at present? eg. daily, weekly, monthly, etc. (if they are never a problem place an 0 next to the problem)

Swallowing difficulties
Choking while eating
Special diet/foods
Abnormal coughing
Chest infections
No control over your wind
Constipation
Diarrhoea (upset stomach)
Smearing in pants
No control over bowels
Wind in abdomen/stomach
No control over bladder
Dribbling urine/water
Burning when passing urine/water
Bladder infections
Problems with sexual functioning
Abdominal/stomach pain

10. How do you presently handle/cope with these problems? eg. what treatments, medications, aids do you use? (if they are never a problem place an 0 next to the problem).

Swallowing difficulties
 Choking while eating
 Special diet/foods
 Abnormal coughing
 Chest infections
 No control over your wind
 Constipation
 Diarrhoea (upset stomach)
 Smearing in pants
 No control over bowels
 Wind in abdomen/stomach
 No control over bladder
 Dribbling urine/water
 Burning when passing urine/water
 Bladder infections
 Problems with sexual functioning
 Abdominal/stomach pain

11. Do you have any other physical problems? (if no, go to question 13)

yes	
no	

12. If yes, describe these

.....

13. Do you have any problems related to sexual functioning? (If you have never had a sexual relationship, go to question 18) (If no, go to question 15)

yes	
no	

14. If yes, describe this

.....

15 Do you experience pain during sexual intercourse?

yes	
no	

16. Have you experienced any problems related to fertility/childbearing?

yes	
no	

17. If yes, what are these

.....

18. Do your physical problems restrict your life in any way? eg. work, sport, socially, etc. (if no, go to question 20)

yes	
no	

19. If yes, describe this

.....

20. Do you have any scars and/or any other physical signs related to your operation? (if no, go to question 22)

yes	
no	

21. If yes, describe these

.....

22. Have any of your family or relations had the same condition as you were born with?
(if no, go to question 24)

yes	
no	

23. If yes, which family members?

.....

24. Do you think you were given enough information/advice about your operation and any associated after-effects/problems? (if no, go to question 2)

yes	
no	

25. If no, what further information would you have liked and at what age would you have needed this?

.....

26. Do you feel you needed any counselling related to any problems you experienced related to your operation and any associated after-effects?
(if no, go to question 28)

yes	
no	

27. If yes, what aspects/topics would you have liked counselling on and at what age would you have needed this?

.....

28. How has your operation and any after-effects affected your life?

.....

29. Describe the most embarrassing incident/happening in your life?

.....
.....
.....
.....

30. Any other comments

.....
.....
.....
.....

Please check to see that you have answered every question. Thank you again for your help.

PAST INFORMATION: (0-20 years)

1 HOSPITALIZATION

1.1 How much time did you spend in hospital when you were growing up as a result of the illness you had when you were born (use probes eg. weeks, months etc., at what age)

.....

.....

.....

1.2 How many operations have you had?
(associated with the illness you were born with)

.....

1.3 Do you think the time you spent in hospital had an effect on your life?

YES
NO

1.4 If yes, describe the effect (use probes eg. relat. with parents esp. mother, schooling, friends, recreation/sport)

.....

.....

.....

.....

.....

2 FAMILY

2.1 Describe the effect that you think your illness had on your family

.....

2.2 Did your illness cause any of the following difficulties:

- financial strain for the family
- restricted family activities
- affected parent/s spare time
- affected relationship between you and your mother
- affected relationship between you and your father
- affected relationship between you and your brother/s, sister/s
- affected your parents relationship

2.3 How did you feel toward your mother and father when you were growing up?

- Very close, could talk about all things
- Fairly close, could talk about most things
- Somewhat close, could talk about some things
- Not very close, could talk about little
- He/she did not live in your house when you were growing up

M

F

3 SCHOOL

3.1 Did you attend a special school at any time during your life?

YES
NO

3.2 If yes, describe this

3.3 Do you think your illness affected your school work?

YES
NO

3.4 If yes, describe this effect

3.5 How often were you absent from school because you had to stay in hospital

3.6 How often were you absent from school because of hospital/clinic /doctor visits?

3.7 How often were you absent from school because of your illness?

3.8 Have you failed a standard or standards at school?

YES
NO

3.9 If yes, which standards did you fail?

3.10 Was your failing associated with your illness

YES
NO

3.11 If yes, explain why

3.12 Did you have any embarrassing incident/s associated with your illness at school? (use probes)

YES
NO

3.13 If yes, describe this/these

3.14 Any other comments (associated with the effect of your illness on your schooling)

.....
.....

4 FRIENDS

4.1 How many close friends did you have (approx.) when you were a child?

4.2 How many close friends did you have (approx.) when you were a teenager?

4.3 Did you have difficulty in making friends?

YES
NO

4.4 If yes, what was the reason?

.....
.....

4.5 Do you think that your illness affected the number of friends that you had when you were growing up?

YES
NO

4.6 If yes, explain why

.....
.....

4.7 Any other comments

.....
.....
.....

5 RECREATION / SOCIAL LIFE

5.1 Did your illness restrict any of your play or sporting activities, when you were growing up?

YES
NO

5.2 If yes, describe in which way were they restricted?

.....
.....
.....

5.3 Did your illness prevent you from staying over at friends?

YES
NO

5.4 If yes, describe why

.....
.....

5.5 Did your illness prevent you from going away on holiday?

YES
NO

5.6 If yes, describe why

.....
.....

5.7 Any other comments

.....
.....
.....

6 DATING

6.1 Did you go out on dates when you were a teenager?

YES
NO

6.2 If yes, how old were you when you had your first date?

.....

6.3 Approx. how many people did you date when you were a teenager?

.....

6.4 Did you have any serious relationships when you were a teenager?

YES
NO

6.5 Do you think that your illness affected your
relationship in any way?

YES
NO

6.6 If yes, in what way?

.....
.....
.....

6.7 Did you have any sexual relationships when you were
a teenager?

YES
NO

6.8 Do you think that your illness affected whether you did, or didn't
have, a sexual relationship?

YES
NO

6.9 If yes, in what way?

.....
.....

6.10 Any other comments

.....
.....
.....

7 MENTAL HEALTH

7.1 How did having your illness, make you feel?

.....
.....

7.2 When growing up, how often did you feel depressed?

- Very often
- Often
- Occasionally
- Rarely
- Never

7.3 When growing up, how often did you feel anxious?

- Very often
- Often
- Occasionally
- Rarely
- Never

7.4 When growing up, how did you feel about your appearance?

- Very satisfied
- Satisfied
- Dissatisfied
- Very dissatisfied

7.5 When growing up, did you have sleeping difficulties?

YES
NO

7.6 If yes, describe these

.....
.....

7.7 When growing up, how frequently did you smoke cigarettes?

- Very often
- Often
- Occasionally
- Rarely
- Never

7.8 When growing up, how frequently did you use alcohol, dagga or similar substances?

- Very often
- Often
- Occasionally
- Rarely
- Never

7.9 When growing up, did you have any weight problems?

YES
NO

7.10 If yes, describe them

.....
.....

PRESENT INFORMATION (over 20 years)

1. POST SCHOOL EDUCATION AND OCCUPATIONAL INFORMATION

1.1 What is your present occupation?

1.2 If you are unemployed, how long have you been unemployed? (In months)

Students and Past Students (Those with no post school qualifications, go to question 2)

1.3 Did your illness affect the choice you made in studying your course/courses?

YES
NO

1.3.1 If yes, in what way
.....

1.4 Did your illness have any effect on your studying?

YES
NO

1.4.1 If yes, in what way
.....
.....

Military Service (If not applicable, go to question 2)

1.5 What is your military service record?

- Completed military service
- Completing military service
- Still to do military service
- Exempted for physical reasons related to illness
- Exempted for other physical reasons
- Exempted for other reasons
- Other

1.6 If you are completing or have completed military service, what is/was your medical classification?

- G1K1
- G3K1
- G3K2
- G3K3
- G3K4
- G4K1
- G4K2
- G4K3
- G4K4
- G7

1.7 Has your illness had any effect on your military service?

YES
NO

1.7.1 If yes, explain why
.....
.....

1.8 Did you experience any embarrassing incidents related to your illness while doing your military service?

YES
NO

☐
☐

1.8.1 If yes, describe them

.....
.....
.....

1.9 Did your difficulties cause any problems between you and other servicemen/superiours?

YES
NO

☐
☐

2. WORK HISTORY

2.1 Describe your work history (use probes eg. number of jobs, length of time etc.)

.....
.....

2.2 Are you presently employed?

YES
NO

☐
☐

2.3 Have you ever been unemployed before?

- Never
- Once
- Twice
- Three times
- More than three times

☐
☐
☐
☐
☐

2.4 For how long were you unemployed each time

.....

2.5 Do you work part time or full time?

PART TIME
FULL TIME

☐
☐

2.6 Has your illness caused any absences from work?

YES
NO

☐
☐

2.6.1 If yes, how often does this occur

.....

2.7 How many days were you absent from work last year?

2.8 For what reason/s were you absent from work

.....
.....

2.9 Have you experienced any embarrassing incidents at work related to your illness?

YES
NO

☐
☐

2.9.1 If yes, describe them

2.10 Do/did your illness cause any problems with those with whom you work?

YES
NO

☐
☐

2.10.1 If yes, describe this

2.11 Did your illness affect your choice of occupation?

YES
NO

☐
☐

2.11.1 If yes, in what way

2.12 Are you satisfied in your present job?

YES
NO

☐
☐

2.12.1 If not, explain why

2.13 Do you think your illness has had any effect on how you do your job?

YES
NO

☐
☐

2.13.1 If yes, explain your answer

3. PEER RELATIONSHIPS

3.1 How many close friends do you have?

None
1
2 - 5
6 - 10
>10

3.2 When last did you see a close friend?

During the previous day
During the previous week
During the previous 2 weeks
During the previous month
More than one month ago

3.3 Do you have difficulty making friends?

YES
NO

3.3.1 If yes, explain why
.....
.....

3.4 How much have you told your friends about your illness?

.....
.....

3.5 Do you presently experience any embarrassing incidents associated with your illness when you are with your friends?

YES
NO

3.5.1 If yes, describe this/these
.....
.....

3.6 Do you think your illness affects the relationship you have with your friends?

YES
NO

3.6.1 If yes, in what way?
.....
.....

3.7 Any other comments
.....
.....

4 MARITAL RELATIONSHIP

4.1 Is this your first marriage?

YES
NO

4.1.1 If no, how many times have you been married before?

4.2 Do you think your illness affected your previous marriage/s?

YES
NO
N/A

4.2.1 If yes, explain why
.....
.....

4.3 Do you think your illness is affecting your present marriage?

YES
NO

4.3.1 If yes, in what way
.....
.....

4.4 Have you told your husband/wife about your illness?

YES
NO

4.5 How would you view your marriage?

Very poor
Poor
Good
Very good

4.6 What is communication like in your marriage?

Very poor
Poor
Good
Very good

4.7 What is honesty like in your marriage?

Very poor
Poor
Good
Very good

4.8 If you have a problem relating to your illness, would you be able to discuss it with your husband/wife?

YES
NO

4.8.1 If no, why not

4.9 Do you think your marriage would be different if you had not been born with your illness?

YES
NO

4.9.1 If yes, explain this
.....
.....

4.10 Any other comments
.....
.....

5 DATING (If not married)

5.1 Do you go out on dates at present?

YES
NO

5.2 Since the age of 20, approximately how many steady relationships have you had?

None
1
2
3
>3

5.3 Do you think your illness affects your current dating/relationships?

YES
NO

5.3.1 If yes, in what way?
.....
.....

5.4 Are you having a steady or serious relationship with anyone at present?

YES
NO

(If no, go to question 5.11)

5.4.1 If yes, how long (in months) have you been "going steady"?

5.5 Do you think your illness is affecting your present steady relationship?

YES
NO

5.5.1 If yes, explain why
.....
.....

5.6 Have you told your partner about your illness?

YES
NO

5.7 How do you view your relationship?

Very poor
Poor
Good
Very good

5.8 What is communication like in your relationship?

Very poor
Poor
Good
Very good

5.9 What is honesty like in your relationship?

Very poor
Poor
Good
Very good

5.10 If you have a problem relating to your illness, could you discuss it with your partner?

YES
NO

5.11 Has telling any of your partners/dates about your illness affected your relationship with them?

YES
NO

5.11.1 If yes, in what way
.....
.....

5.12 Do you think you would have had more dates or serious/steady relationships if you had not been born with your illness?

YES
NO

5.12.1 If yes explain why
.....
.....

5.13 Any other comments
.....
.....

6 SEXUAL RELATIONSHIP/S

6.1 Have you had any sexual relationships since you were a teenager?

YES
NO

6.2 If yes, do you think your illness had any effect on these relationships?

YES
NO

6.2.1 If yes, in what way?
.....
.....

6.2.2 If no, do you think your illness affected your not having sexual relationships?

YES
NO

6.2.2.1 If yes, why?
.....
.....

6.3 Are you having a sexual relationship at present?

YES
NO

6.3.1 If yes, how would you view this relationship?

Very poor
Poor
Good
Very good

6.4 Does your illness affect your view of how sexually attractive you are to others?

YES
NO

6.5 If yes, explain why
.....
.....

6.6 Does your illness affect your sexual functioning?

YES
NO
N/A

MALES ONLY

6.7 Do you have any erection problems? (eg. obtaining an erection, maintaining an erection, semi-erections etc.)

YES
NO

6.7.1 If yes, describe this

6.8 Do you have problems with ejaculation? (eg. no ejaculation, premature ejaculation)

YES
NO

6.8.1 If yes, describe this?

6.9 If you do experience any of the above difficulties, are they related to your operation or illness?

YES
NO

6.10 If yes, explain your answer

FEMALES ONLY

6.11 Do you suffer from vaginismus?

YES
NO
N/A

6.12 Do you have any orgasmic problems?

YES
NO
N/A

6.12.1 If yes, describe this

6.13 If you do experience any of the above difficulties, are they related to your operation or illness?

YES
NO

6.13.1 If yes, explain your answer

MALES AND FEMALES

6.14 Do you think your sexual experiences/relationships would have been different if you had not been born with your illness?

YES

NO

6.14.1 If yes, explain your answer

6.15 Any other comments

7 MENTAL HEALTH

7.1 How do you feel, at present, about life as a whole?

Very satisfied
Satisfied
Dissatisfied
Very dissatisfied

7.2 How do feel about yourself in general? (eg. Do you like yourself)

Very satisfied
Satisfied
Dissatisfied
Very dissatisfied

7.3 How do you feel about your appearance in general?

Very satisfied
Satisfied
Dissatisfied
Very dissatisfied

7.4 What part of your body do you dislike most?

7.5 At present, how often do you feel depressed?

Very often
Often
Occasionally
Rarely
Never

7.6 At present, how often do you feel anxious?

Very often
Often
Occasionally
Rarely
Never

7.7 Do you find it difficult to fall asleep

Very often
Often
Occasionally
Rarely
Never

7.8 How often do you smoke cigarettes?

Very often
Often
Occasionally
Rarely
Never

7.9 How frequently do you use dagga or similar substances?

Very often
Often
Occasionally
Rarely
Never

7.10 How often do you use alcohol?

Very often
Often
Occasionally
Rarely
Never

7.11 When you are anxious or upset do you eat more than usual?

Very often
Often
Occasionally
Rarely
Never

7.12 When you are anxious or upset do you eat less than usual?

Very often
Often
Occasionally
Rarely
Never

7.13 Have you ever served a jail sentence?

YES
NO

7.14 If yes, descibe this
.....
.....

7.15 Do you like mixing with people?

- Very often
- Often
- Occasionally
- Rarely
- Never

7.16 Do you think you think you have any psychological difficulties at present? (eg. severe depression, abnormally aggressive feelings)

- YES
- NO

7.17 If yes, describe this/these
.....
.....
.....

7.18 At present what upsets you most about your illness?
.....
.....
.....

7.19 Are any of the following feelings or behaviour mainly associated with your illness?

- Self esteem (How you feel about yourself)
- Body image (How you feel about your body)
- Anxious feelings
- Sleeping difficulties
- Cigarette smoking
- Drug use
- Alcohol use
- Overeating
- Under eating
- Criminal offences
- Mixing socially
- Depression
- Psychological difficulties

7.20 Any other comments
.....
.....
.....

8. SOCIAL LIFE/RECREATION

8.1 How would you rate your social life at present?

Very poor
Poor
Good
Very good

8.2 What do you spend most of your spare time doing?
.....
.....
.....

8.3 To how many clubs/social organizations do you belong?

None
1
2
3
>3

8.4 Do you participate in any sport?

YES
NO

8.4.1 If yes, name them

8.5 Does your illness restrict social and/or sports activities.

YES
NO

8.5.1 If yes, in what way?
.....
.....

8.6 Do you prefer being alone or with others in your spare time?

Alone
With others

8.7 Are there some recreational/social/sports activities that your friends take part in, that you can't?

YES
NO

8.7.1 If yes, which ones?
.....

8.8 Any other comments
.....
.....
.....

9 MEDICAL AND SOCIAL WORK SERVICES

9.1 Medical Service

9.1.1 How do you feel about the quality of the treatment you received at this hospital? (use probes eg. good, bad).....

9.1.2 How do you feel about the success of the operations you had at this hospital?

9.1.3 How do you feel about medical treatment you received from the doctors?

9.1.4 How do you feel about medical treatment you received from the nurses?

9.1.5 From who else did you receive treatment at this hospital ?(eg. stomatherapist, physiotherapist, etc.)

9.1.6 What do you think of the treatment you recieved as an inpatient?

9.1.7 At what age were you first told about your illness and operation/s?

9.1.8 Who told you?

9.1.9 Are you satisfied with the amount of information that the doctors gave you concerning your illness?

YES

NO

9.1.9.1 If no, why not

9.1.10 Are you satisfied with the way that the doctors, nurses and other hospital staff treated you? (Besides medical treatment).

YES

NO

9.1.10.1 If no, why not?

9.1.11 Up to what age did you attend this hospital?

9.1.12 If you did not come back to this hospital after the age of 14, how do you think you would have felt, had you come back for treatment?
.....
.....

9.1.13 Did you attend another hospital, clinic or go to other doctors after attending this hospital?

YES
NO

9.1.13.1 If yes, describe this
.....
.....

9.1.13.2 If yes, how did you feel about this change? (eg. what was it like seeing new doctors etc.)
.....

9.1.14 Do you think there is a need for a clinic or hospital specifically for teenagers born with your, or other illnesses?

YES
NO

9.1.14.1 Explain your answer
.....
.....

9.1.15 In what way/s do you think the medical service at this hospital could be improved?
.....
.....

9.1.16 In what other ways do you think medical service for people born with your illness could be improved? (eg. facilities, resources in the community)
.....
.....

9.1.17 Overall, do you remember your contact with this hospital as being positive or negative?

Positive
Negative

9.1 17.1 Explain your answer
.....

9.1 18 Any other comments
.....

9.2 Social Work Services

9.2.1 Have you received any professional counselling eg. psychologist, socialworker, psychiatrist?

YES
NO

☐
☐

9.2.1.1 If yes, from whom did you receive counselling?

9.2.2 Do you think you ever needed professional help/counselling?

YES
NO

☐
☐

9.2.2.1 If yes, explain why
.....

9.2.3 Do you think you need professional help/counselling now?

YES
NO

☐
☐

9.2.3.1 If yes, explain why?
.....

9.2.4 At what age/s did you need help most?

9.2.5 What topics/issues did you need to discuss?
.....

9.2.6 Did you ever receive any counselling from this hospital ie. from a social worker, psychiatrist, psychologist, stomatherapist, any other hospital staff?

YES
NO

☐
☐

9.2.6.1 If yes, describe this
.....

9.2.7 Do you think that people born with your illness need professional counselling?

YES
NO

☐
☐

9.2.7.1 If yes, explain why
.....
.....

9.2.8 In what way do you think the social work service at this hospital could be improved?
.....
.....

9.2.9 In what other ways do you think the social work/psychological services for people born with your illness could be improved ie. facilities, resources in the community?
.....
.....

9.2.10 Any other suggestions concerning social work services
.....

9.2.10 Any other comments
.....

10 EVALUATION

10.1 How has answering this questionnaire made you feel? (Anxious, angry etc.)
.....
.....
.....
.....
.....

APPENDIX 6

BATSON SCORING SYSTEM

Counting any person of 10 years and over as one equivalent person, and any person under ten years as half an equivalent person. The number of rooms needed is as follows:

Up to 2,5 equivalent persons	1 room
3,5	2 rooms
5,0	3 rooms
7,5	4 rooms
10,0	5 rooms
12,5	6 rooms
15,0	7 rooms
17,5	8 rooms
20,0	9 rooms

Consider only the rooms used for sleeping in the household

Formula:

Number of equivalent persons in house x 100

Number of persons allowed per rooms 1
used for sleeping

Percentage of overcrowding

100% = uncrowded

100% - 149% = overcrowded

100% = crowded

150% = gross overcrowding

(Batson 1944)

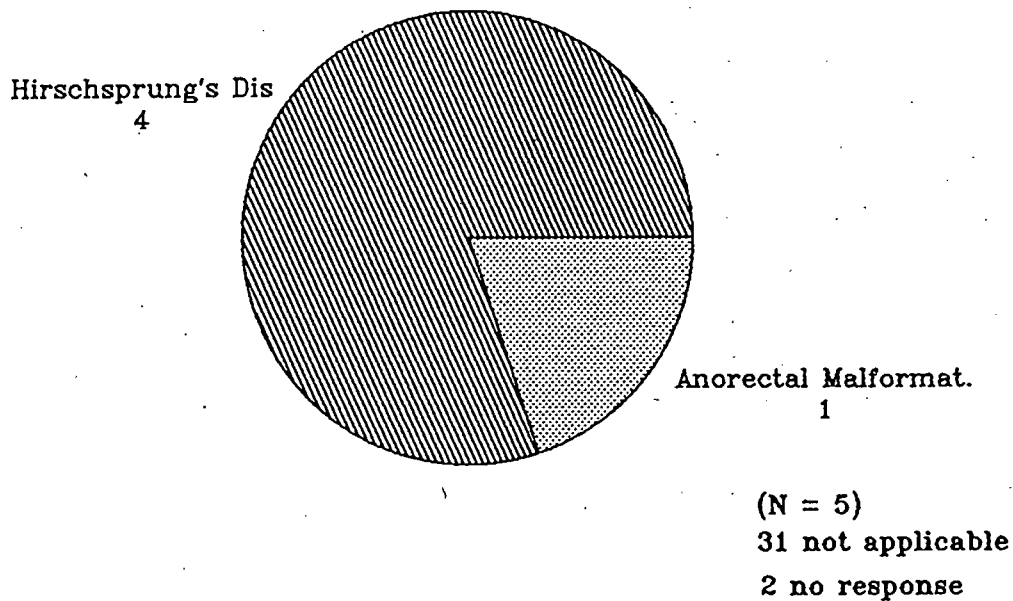
APPENDIX 7

CLASSIFICATION OF OCCUPATIONS

Social class	Type of occupation
I	Higher professional
II	Other professional and technical
III (non-manual)	Other non-manual occupations
III (manual)	Skilled manual
IV	Semi-skilled manual
V	Unskilled manual

(Davie, Butler, Goldstein 1972:3)

Diagram 1 DIAGNOSIS BY OPERATIONS
RELATING TO CONGENITAL ANOMALY *



* Since those at the Children's Hospital

Diagram 2 DIAGNOSIS BY PARENTS'
RELIGIOUS ATTENDANCE

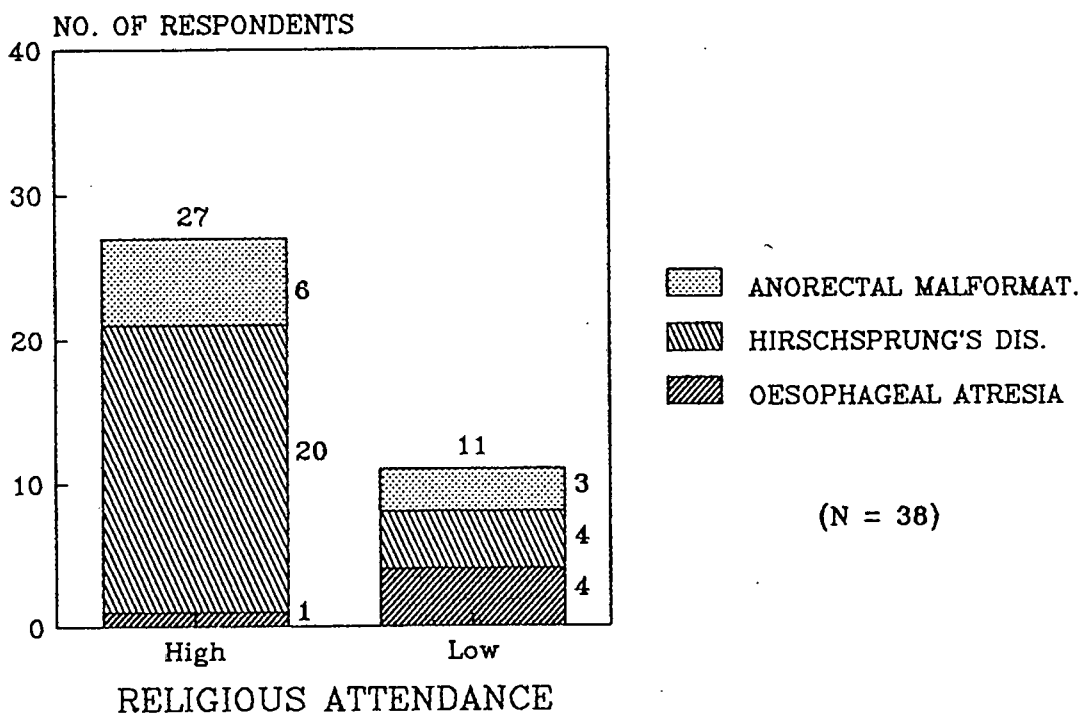
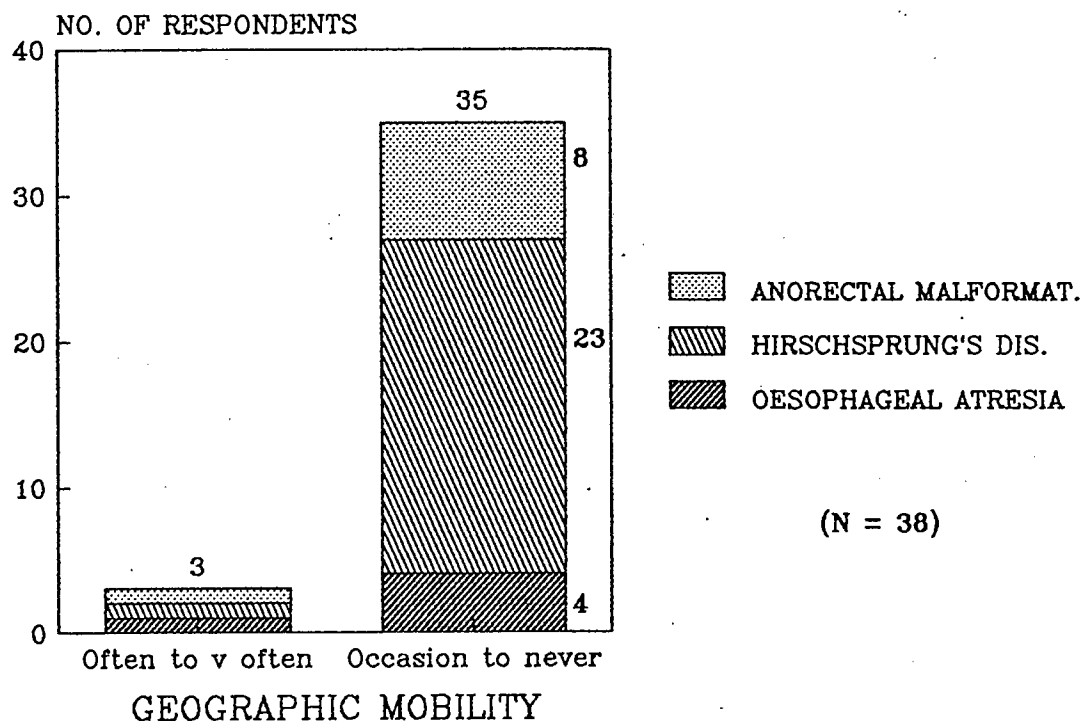


Diagram 3 DIAGNOSIS BY GEOGRAPHICAL MOBILITY *



* Frequency of moving house

Diagram 4 DIAGNOSIS BY STANDARD OF LIVING

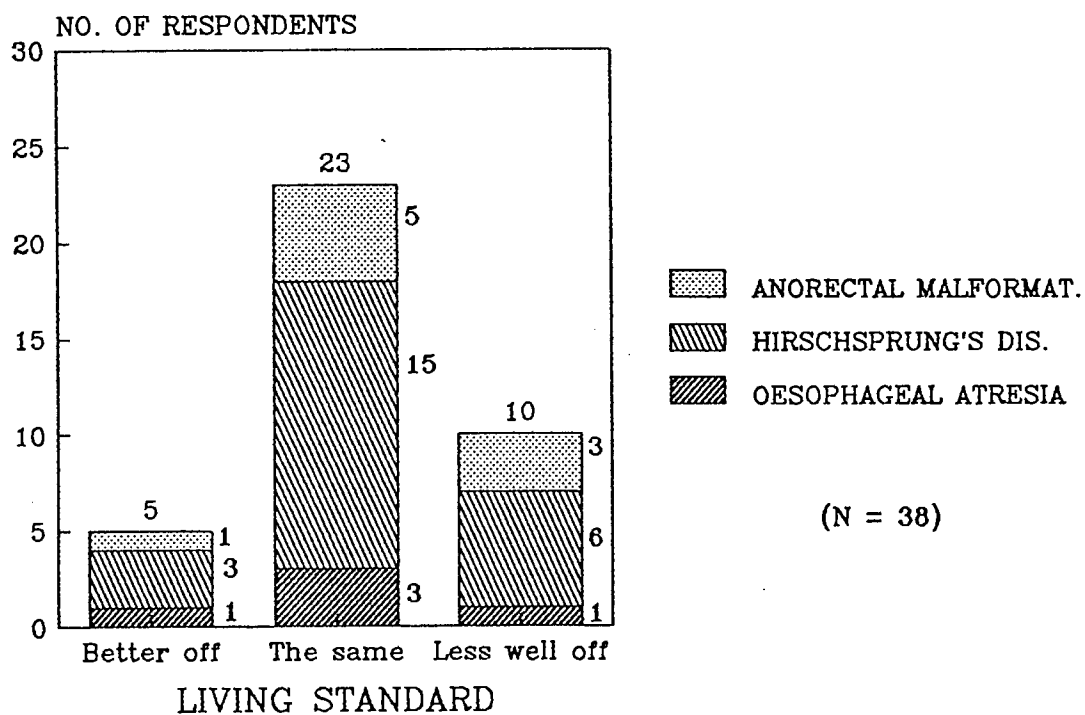


Diagram 5 DIAGNOSIS BY LIVING ARRANGEMENTS

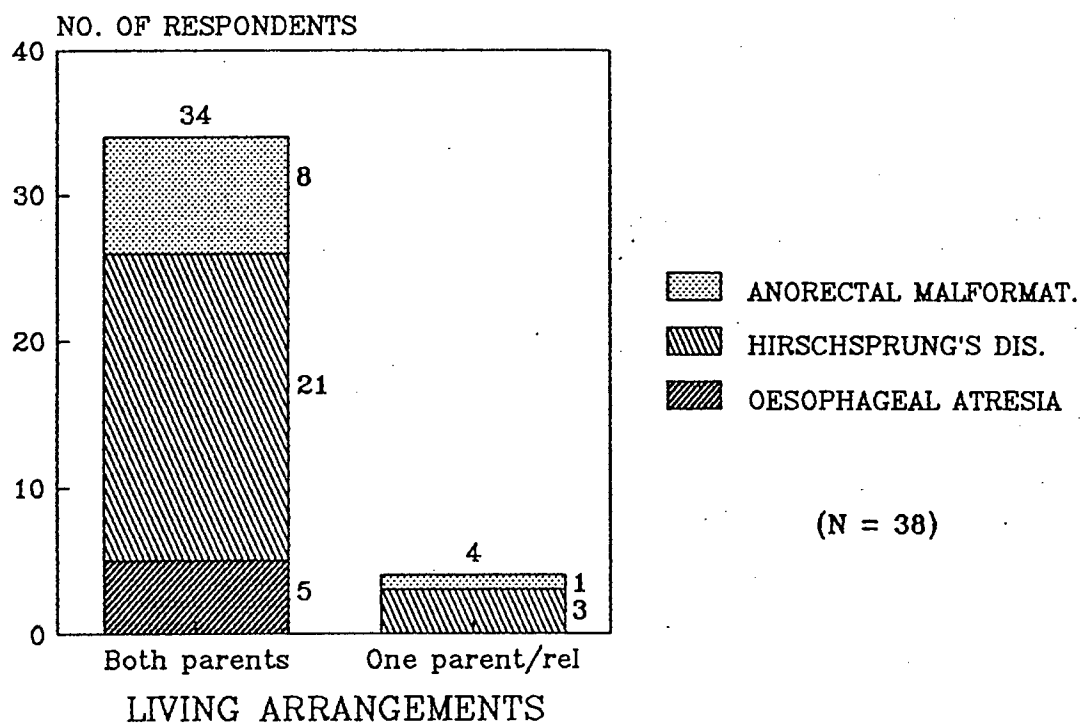


Diagram 6 DIAGNOSIS BY NO. OF SIBLINGS

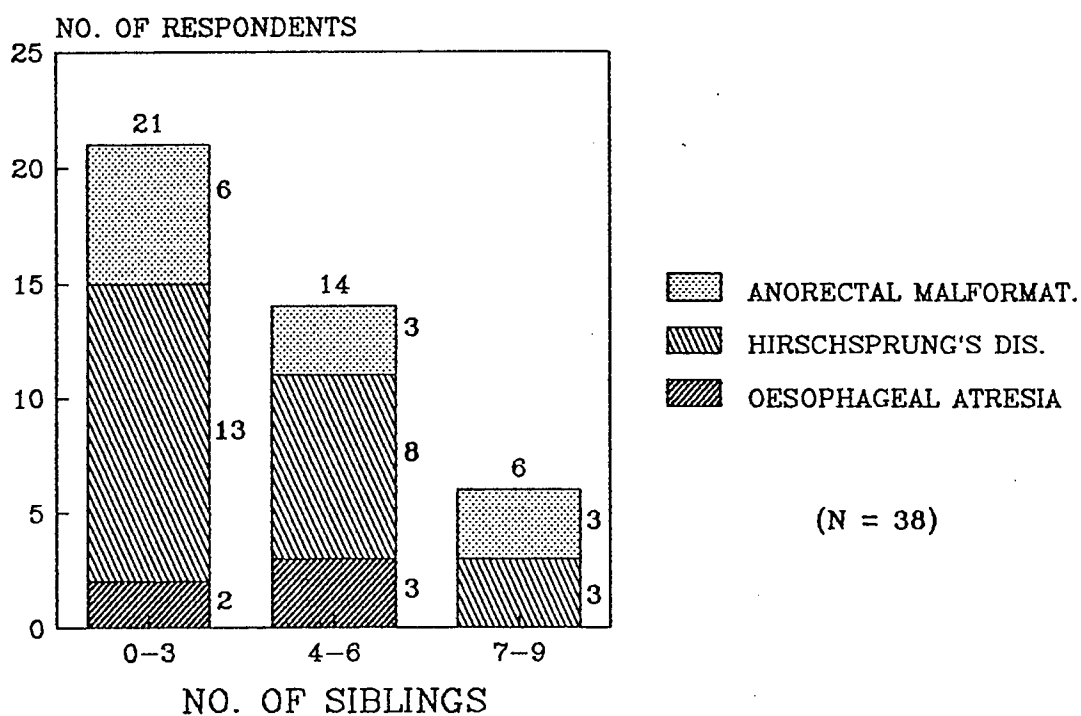


Diagram 7 DIAGNOSIS BY FATHER'S
EMPLOYMENT STATUS

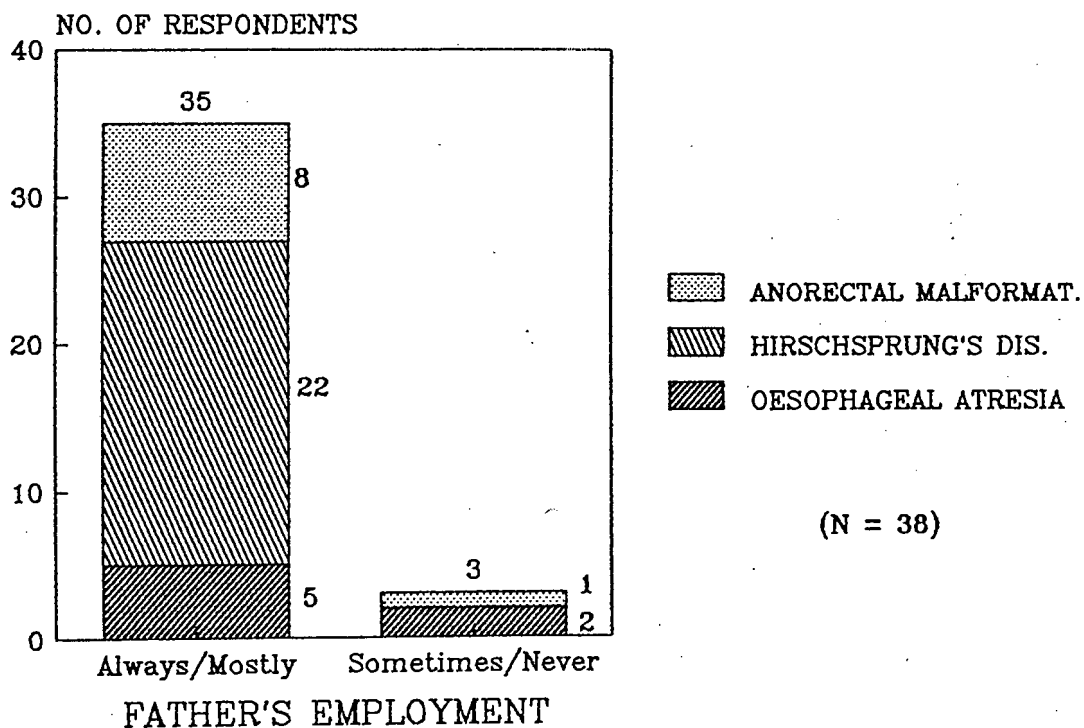


Diagram 8 DIAGNOSIS BY MAIN BREADWINNER

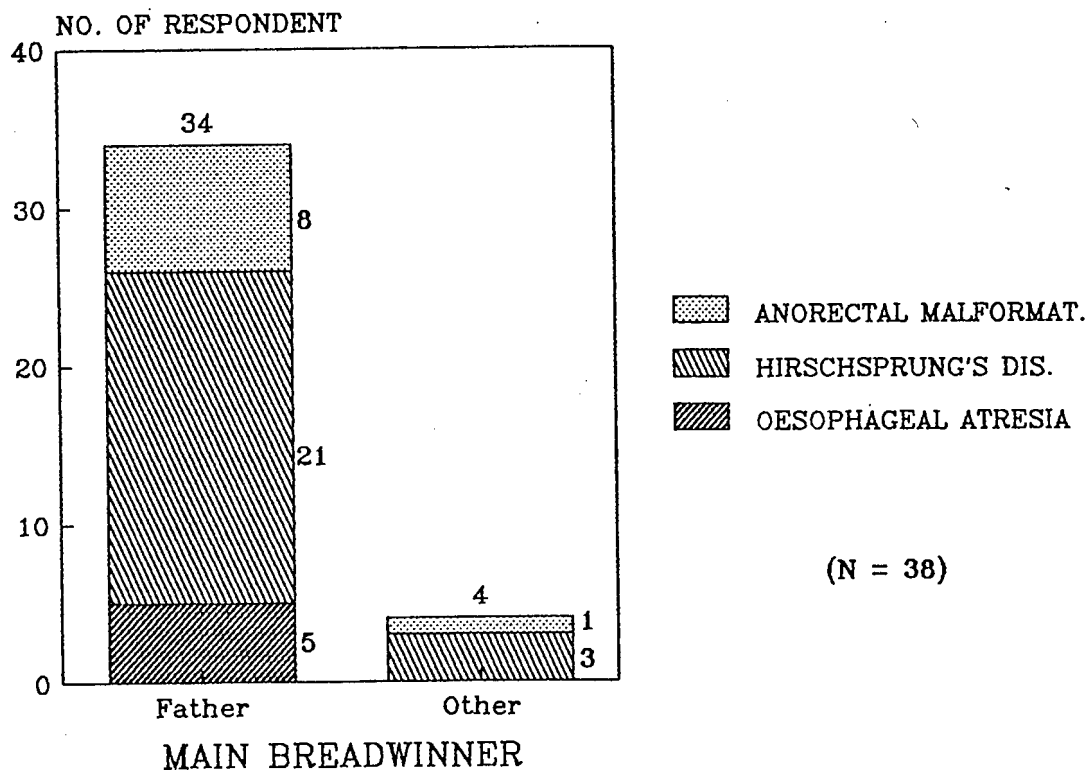


Diagram 9 DIAGNOSIS BY TYPE OF HOUSING

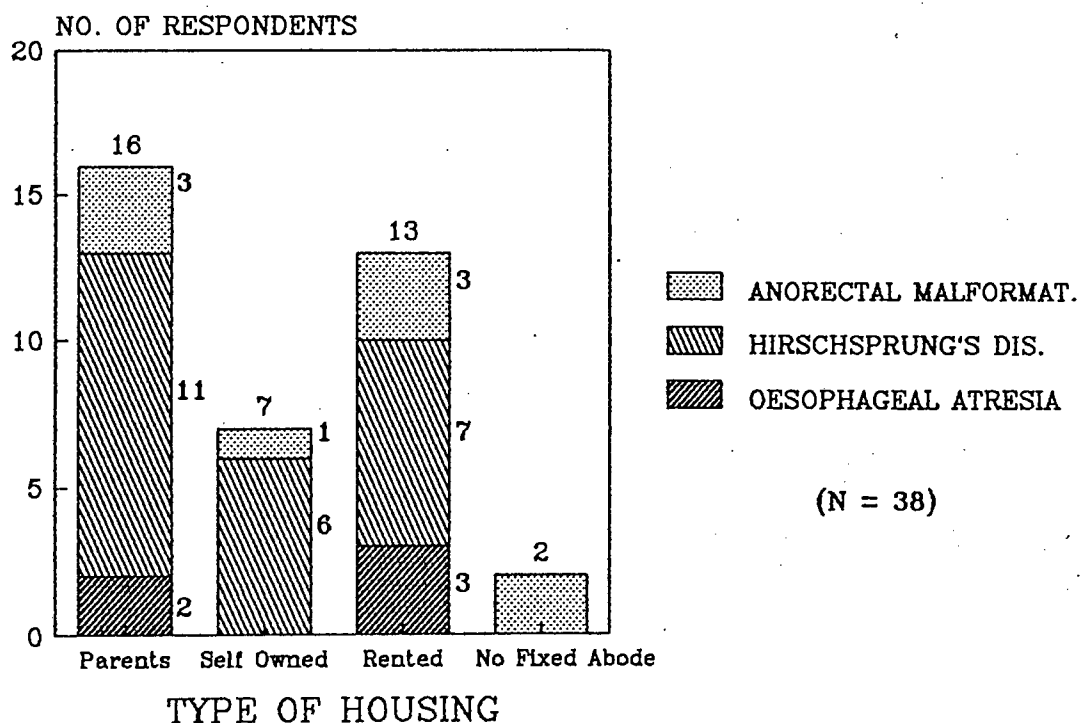
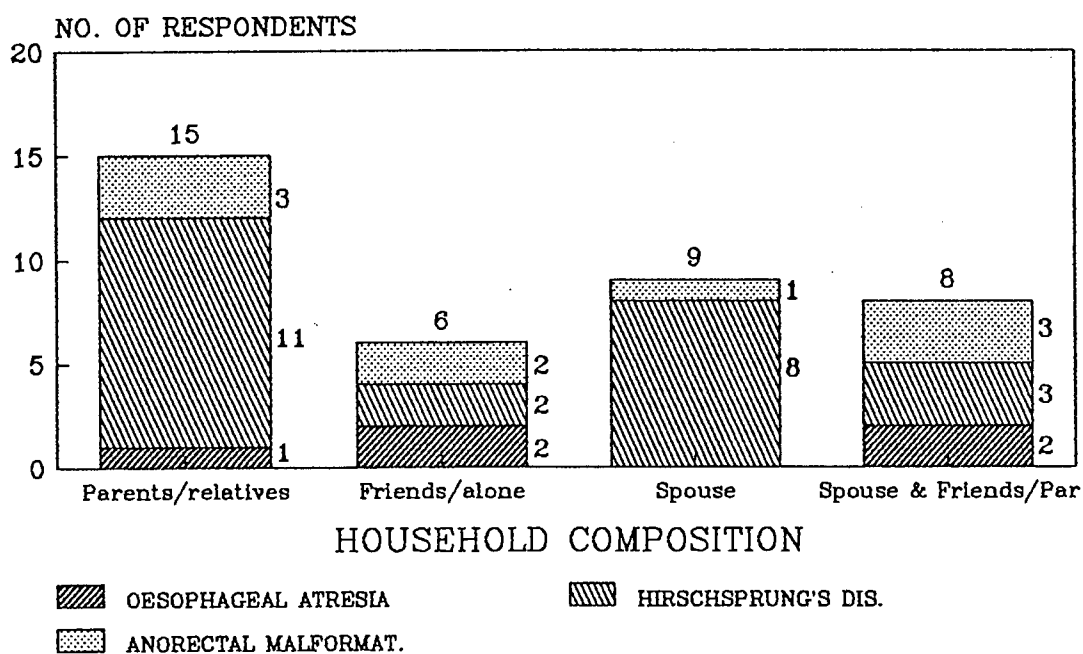


Diagram 10 DIAGNOSIS BY HOUSEHOLD COMPOSITION *



(N = 38)

* People living with the respondent

Diagram 11 DIAGNOSIS BY OVERCROWDING
(OF HOUSING)

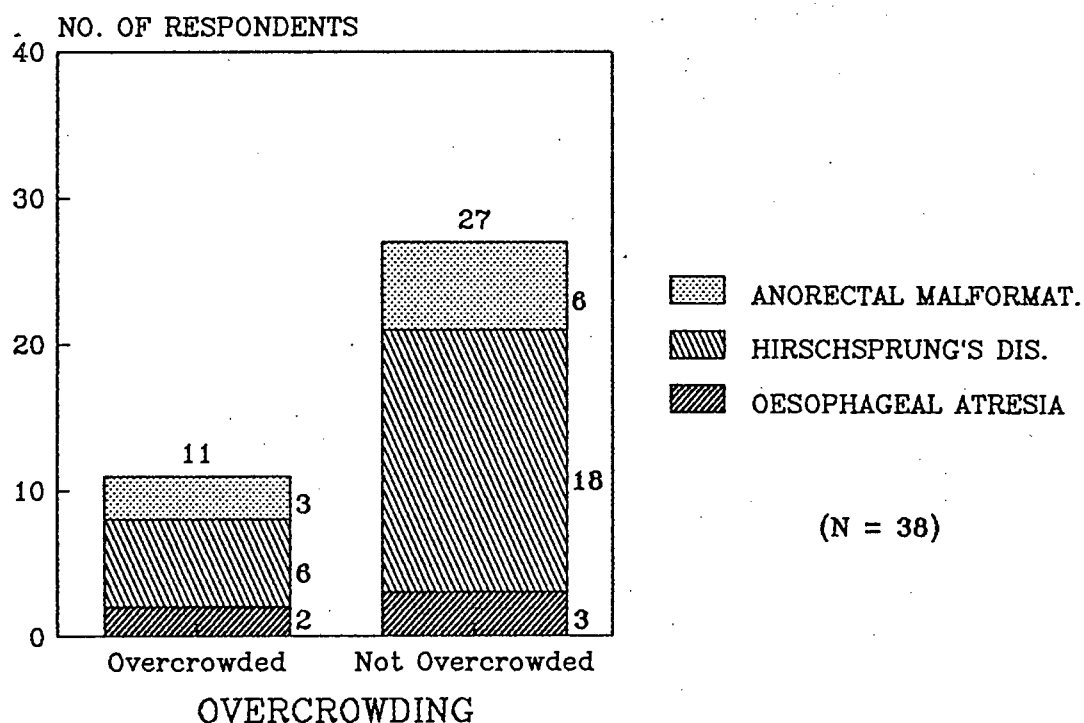


Diagram 12 DIAGNOSIS BY EMPLOYMENT
STATUS

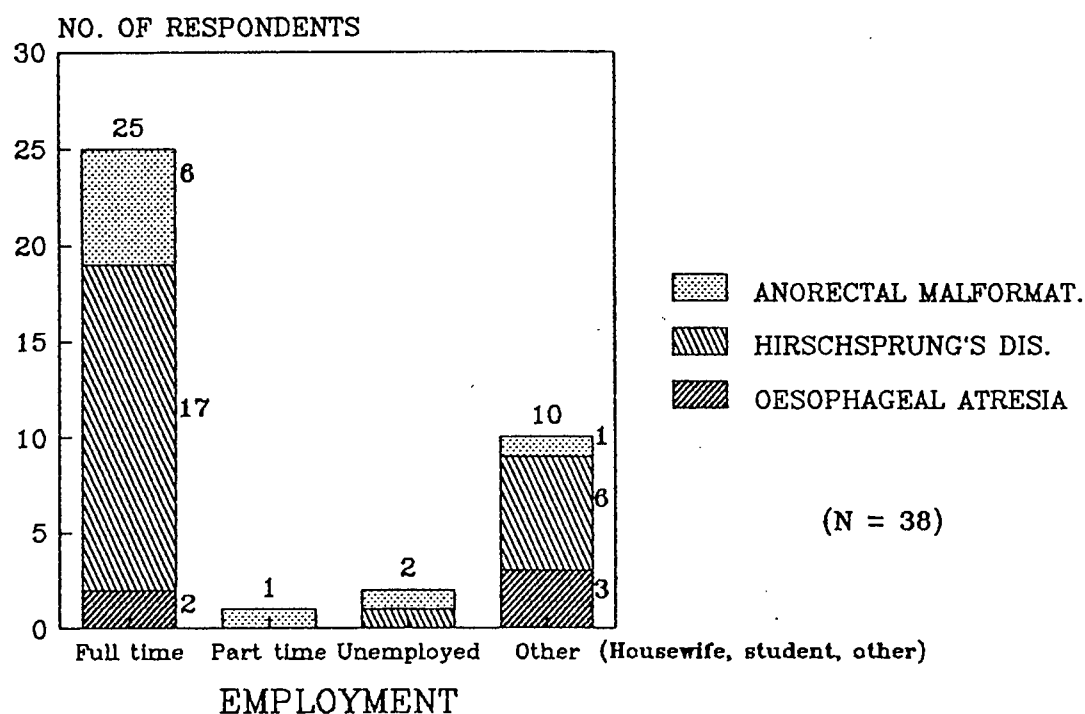


Diagram 13 DIAGNOSIS BY MOTHER'S EDUCATION

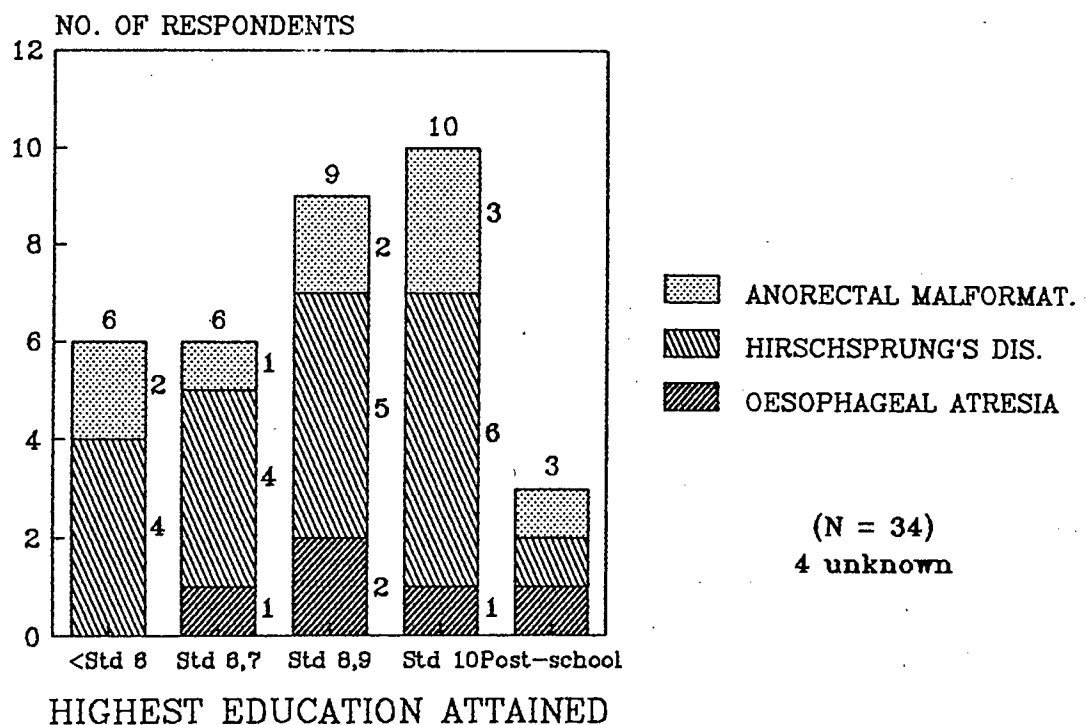


Diagram 14 DIAGNOSIS BY PARENT DEATH/ILLNESS

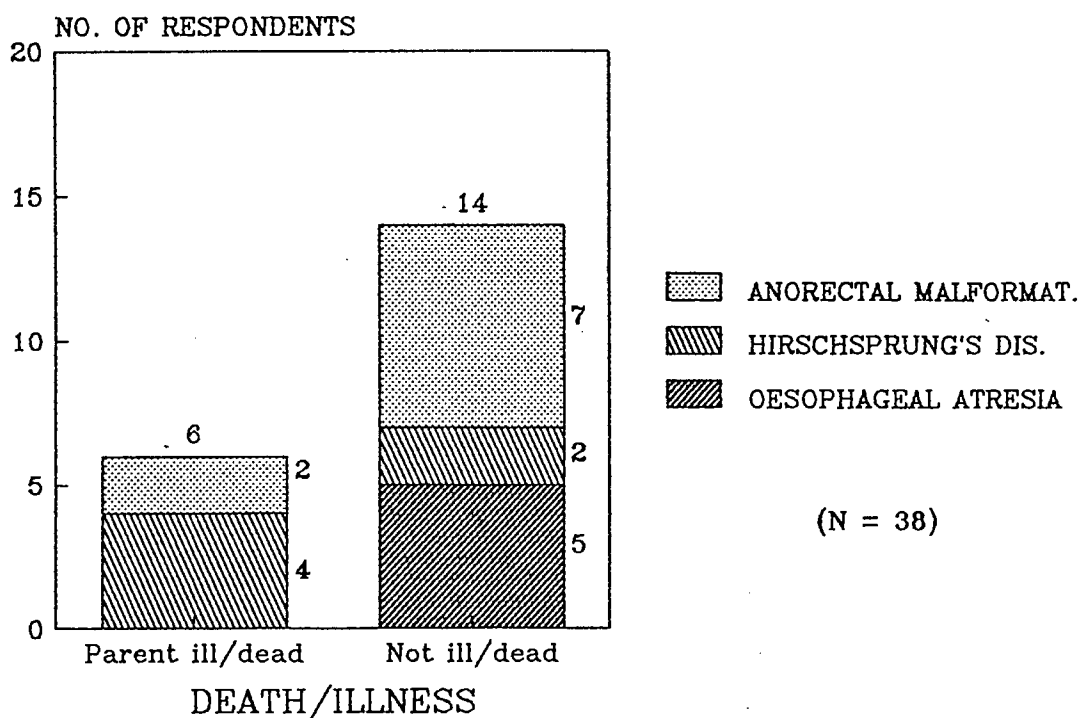


Diagram 15 DIAGNOSIS BY MARITAL PROBLEMS

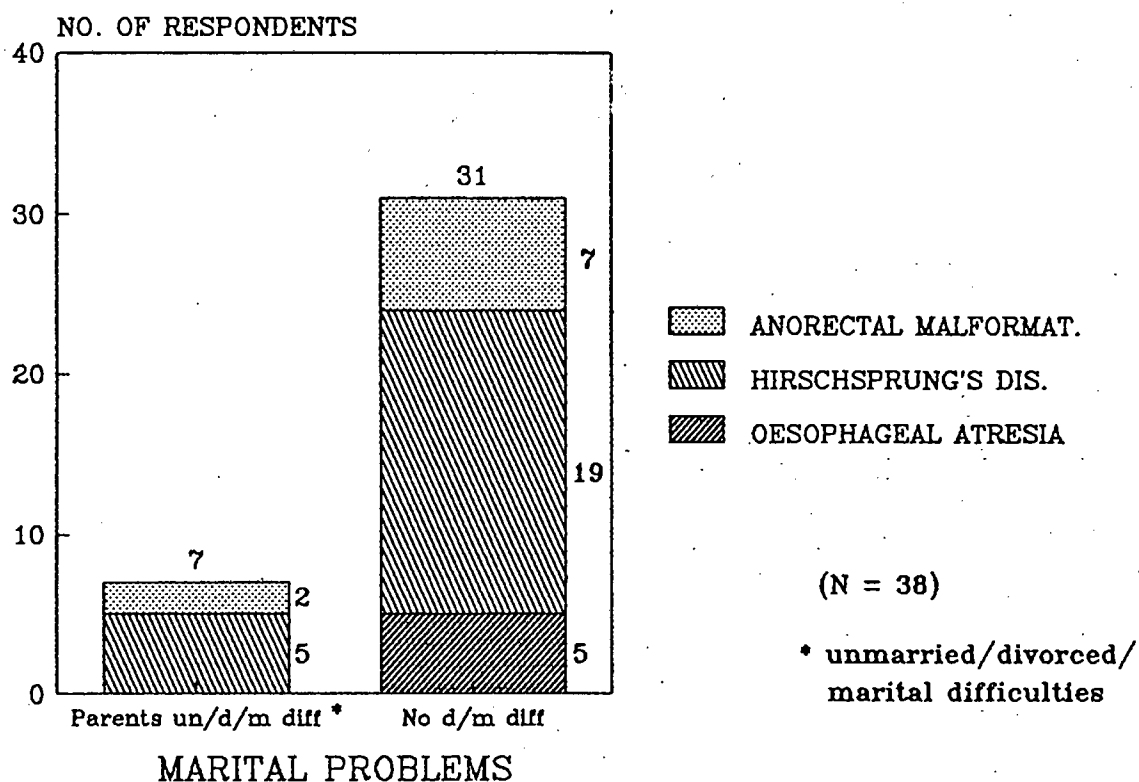


Diagram 16 DIAGNOSIS BY PARENTS' PSYCHIATRIC PROBLEMS

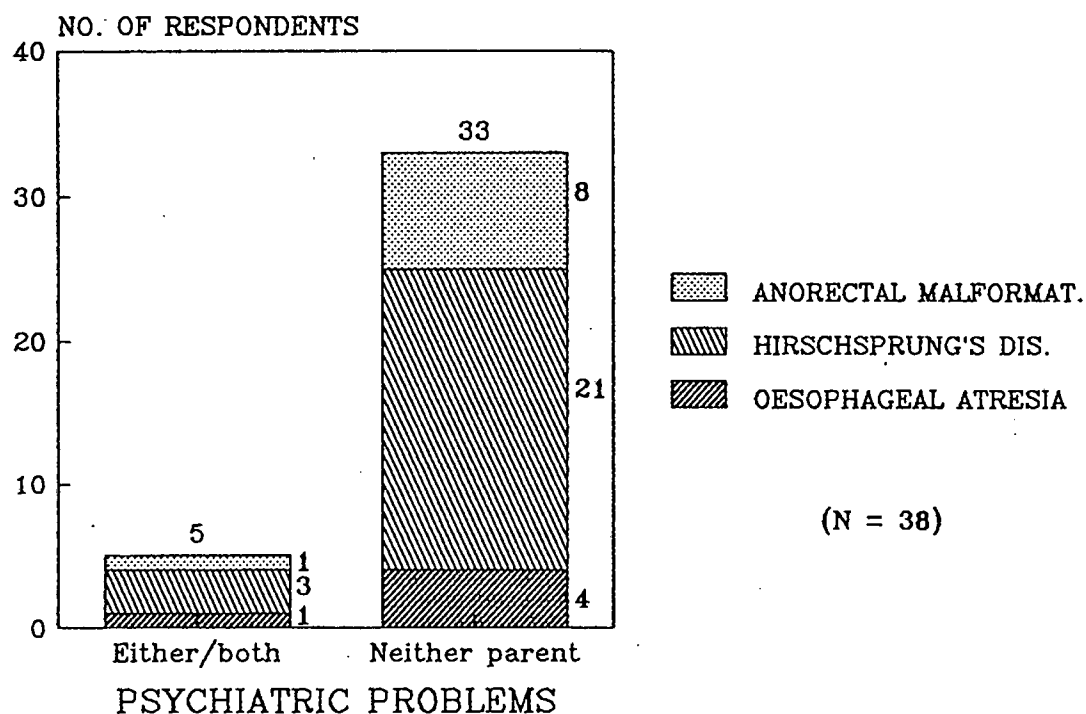


Diagram 17 DIAGNOSIS BY PARENTS'
CRIMINAL OFFENCES

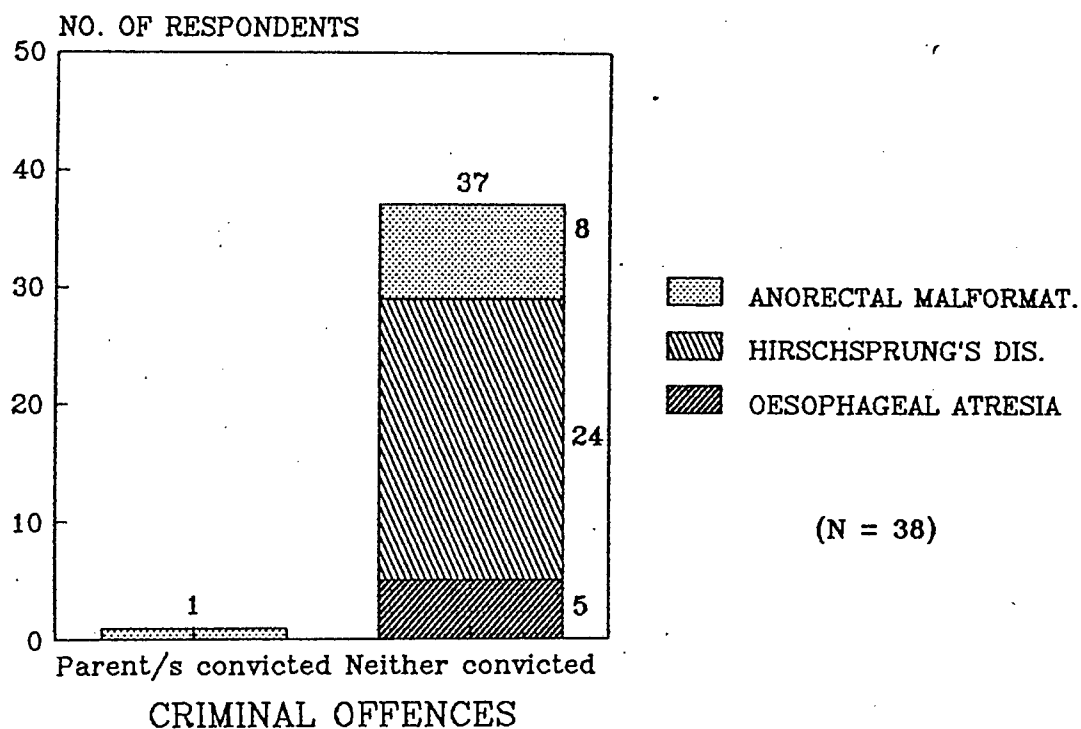


Diagram 18 DIAGNOSIS BY PARENTS
SUBSTANCE ABUSE

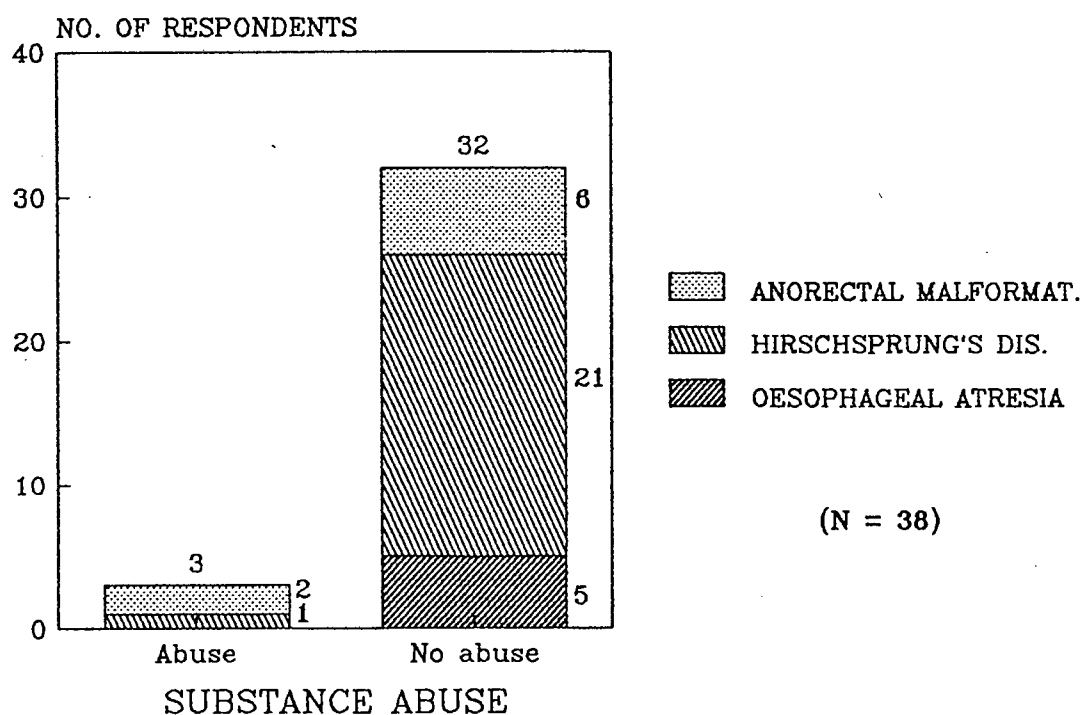
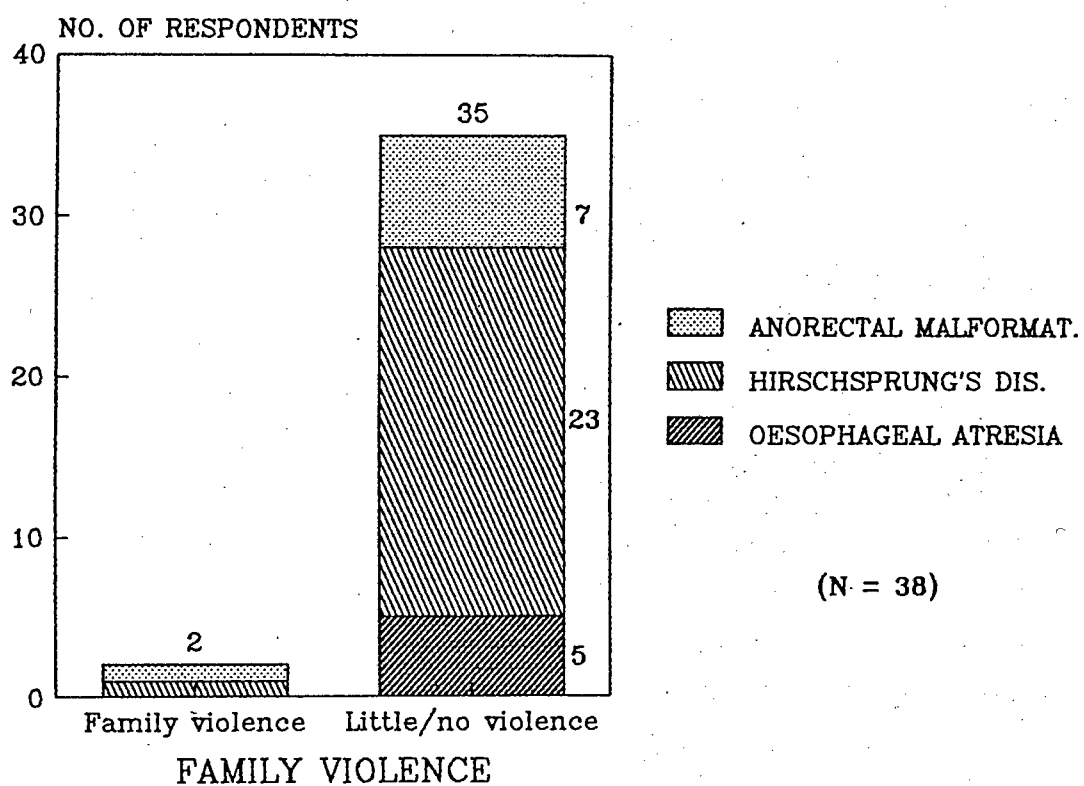


Diagram 19 DIAGNOSIS BY FAMILY VIOLENCE



APPENDIX 9

RESIDUAL PROBLEMS - ALL GROUPS (PAST AND PRESENT)

(d refers to daily; w refers to weekly; m refers to monthly. When questioned on the age at which a problem occurred, some patients wrote 'always' which meant from birth to their present adult life. Some patients referred to the age at which a problem occurred, some patients referred to the frequency with which it occurred and some patients referred to both - this explains discrepancies in responses. Most problematic age refers to age in years.)

A. Oesophageal atresia

(a) Residual problems (past)

(i) Most problematic age and how often they occurred

Swallowing difficulties - 0-4(d); 0-15(w); 0-15(d).

Choking while eating - 0-4(m) 0-10(w); 0-15(d).

Abnormal coughing - 0-4(m); 0-14(m); 0-30

Chest infection - 0-22(m).

(ii) Age at which residual problems stopped

Swallowing difficulties 14 (2 cases are still a problem)

Choking while eating (3 are still a problem)

Abnormal coughing (2 are still a problem)

Chest infections (1 is still a problem).

(iii) Coping/treatment methods

Swallowing difficulties - dilatations (1); constricting throat to force it down (2); avoid cereal and dairy products (1).

Choking while eating - dilatations (1); constricting throat to force food down (1); drinking water (1)

Abnormal coughing - drinking water (1)

Chest infections - medication (1)

(b) Residual problems (present)

(i) How often problems occur

Swallowing difficulties - m(2); d(1)

Choking while eating - m(2); y(1)

Abnormal coughing - m(2)

Chest infections - y(1)

(ii) Coping/treatment methods

Swallowing difficulties - Constrict throat muscles (2)

Choking while eating - Drink water or vomit (1); dilatation

Abnormal coughing - medication (1)

Chest infections - medication (1).

B. Hirschsprung's disease

(a) Residual problems (past)

(i) Most problematic age and how often they occurred

Special diet: 8-10(w); 15-16(d); 6-14(d); always (d); 0-24 (d);
always (d); 1-27(d)

No control over flatus 6(w); always (d); 14(w); always (d);
always (w); always (d); 0-19(d); 0-10(w); 1-2(d)

Constipation always(w); always(d) 5-15(m); 12-13(w); always (w);
13-20(m)

Diarrhoea 6(w); 1-15(w); always(d); 10(w); 5-15(w); always(w);
6-10; 0-10(w); always(d); 12(w); always(d) 0-19(d); 0-10(d);
10-15(m)

Smearing 1-2(d); 8-9(w); 6-10(m); always(d); 0-17(d); always(d);
always(w); 6-10; 0-7(w); always(d); always(d); always(w); 0-19(d)

No control over bowels 1-27(w); 1-15(m); 8-9(m); 6-10(m)
always(d); 0-16(d); 0-10(d); 0-19(d); 0-7(w); 5-15(d); always(d);

0-24(d); 6-18(w); always

Excessive flatus 1-27(d); 6(m); 18(d); 12(d); always (w);
always(d); always; 12-13(w); always(w); 0-19(d); 1-15(w)

No control over bladder 6-10(m)

Dribbling urine 6-10(m); 20-21(m); always

Burning when urinating 20(m); 20-21(m)

Abdominal pain 6(m); 1-15(w); 16-17(d); 0-15; always(d);
12-13(m); always(w); always(w); 0-19(d).

(ii) Age at which residual problem stopped

Special diet 16,24 (5 cases are still a problem)

No control over flatus (8 cases are still a problem)

Constipation 19,16 (6 cases are still a problem)

Diarrhoea 17,10,16 ((8 cases are still a problem)

Smearing 15,10,17,16 (9 cases are still a problem)

No control over bowels 13,10,17,16 (3 cases are still a problem)

Excessive flatus (10 cases are still a problem)

No control over bladder 10,18

Dribbling urine 10 (1 cases still a problem)

Burning when urinating (2 cases still a problem)

Bladder infections 10 (1 case still a problem)

Abdominal pain (8 cases still a problem)

(iii) Coping/treatment methods

Special diet - avoided fatty foods (1); roughage or acidic food
(2); fizzy cooldrinks (2); fruit juices (1); curry (2); tomatoes

No control over wind - medication (1)

Constipation - medication (5); salt water (1); drinking boiling
water (1); Fybrogel (1); Kayomagina (1); Codeine Phosphate (1).

Diarrhoea - flour water (1); Balsam Vitae (2); Chamberlains Colic
(2); Brown or wholewheat bread (1); Immodium (2); Lomotil (1);

Peter Pack (1)

Smearing - wore nappy (2); change underwear often (3); put toilet paper in underwear (1); frequent bowel action (1); Immodium (1)

No control over bowels - wore nappy (1); wore nappy till the age of 14 (1); toilet paper in underwear (1); frequent change of underwear (1); medication (4)

Wind in abdomen - Enos (2); medication (2)

No control over bladder - wore nappy

Dribbling urine - frequent change of underwear

Bladder infections - medication (2)

Abdominal pain - medication (6)

(b) Residual problems (present)

(i) How often problems occur:

Special diet d(3 cases); w(1 case)

No control over flatus d(8); w(2)

Constipation d(3); w(3); m(3)

Diarrhoea d(3); w(5); m(2)

Smearing d(4); w(3); m(1)

No control over bowels w(1); d(1); m(1)

Excessive flatus d(7); w(3); m(2)

Dribbling urine d(1)

Burning when urinating d(1); m(1)

Bladder infections y(1)

Abdominal pain d(2); w(5); m(1)

(ii) Coping/treatment methods

Special diet - avoid fatty foods and acid foods (1)

No control over flatus - Balsam Vitae (1); Regulettes(1); boiling water (1); medication (1); try and hold it in - very painful (1)

Constipation - laxatives (1); medication (1)

Diarrhoea - Peter Pack (1); brown bread (1); medication (1); regular bowel action (1)

Smearing - change underwear frequently (2); toilet paper in underwear (2); regular bowel action

No control over bowels - Balsam Vitae (1); Chamberlain Colic (1)

Excessive flatus - Enos (1); Rennies (1); medication (1); ant-acid (1); regular bowel action (2)

Dribbling urine - change underwear (1)

Abdominal pain - colic medication (1); other medication (2).

C. Anorectal malformations

(a) Residual problems (past)

(i) Most problematic age and how often the problem occurred

Special diet - 7-10(d); 11(w)

No control over flatus - 0-22(d); 0-24(d); 10(w); 0-12(w)

Constipation - 1-28(d); 0-22(d); 0-24(m); 6-12; 0-35(d)

Diarrhoea - 1-6(d); 13(d); 0-24(m); 0-17

Smearing - 1-28(d); 0-22(d); 1-27(d); 0-24(d); 0-17(d); 0-28(d); 0-28(m); 0-12(w)

No control over bowels - 1-28(d); 0-17(d); 0-24(d); 0-17(d);

Excessive flatus - 1-22(d); 0-24(d); 0-23(d); 0-35(w)

No control over bladder - 0-12(d); 0-12(d)

Dribbling urine - 1-18(d); 0-28(d)

Burning when urinating - 0-22(m)

Abdominal pain - 7-14(d); 0-35(w)

(ii) Age at which residual problem stopped

Special diet - 10,20

No control over wind - (3 cases still a problem)

Constipation - 20 (3 cases still a problem)

Diarrhoea - 16,15,20 (2 cases still a problem)

Smearing - 19 (5 cases still a problem)

No control over bowels - 17,19 (3 cases still a problem)

Wind in abdomen - (3 cases still a problem)

No control over bladder - 12

Dribbling urine - 18 (1 case still a problem)

Abdominal pain - (2 cases still a problem)

(iii) Coping/treatment methods

Special diet - avoid beans, cabbage, apples (1), eat potatoes (1)

Constipation - laxatives(2); exercise(1); bananas (1); Senokots (1)

Diarrhoea - self-raising flour (1); brown bread (1); Immodium (1); medication (1)

Smearing - change underwear (2); bowel washouts (1); toilet paper in underwear (2)

No control over bowels - operation (1); colostomy re-opened (1); bowel washouts (1)

Excessive flatus - Enos (1)

Dribbling urine - dry penis after urinating.

(b) Residual problems (present)

(i) How often problems occur

No control over flatus - d(2); w(2)

Constipation - d(1); w(1); m(3)

Diarrhoea - w(1); m(3)

Smearing - d(3); w(1); m(1)

No control over bowels - d(2)

Excessive flatus - d(2)

Dribbling urine - w(1); d(1)

Abdominal pain - w(2); m(1)

(ii) Coping/treatment methods

Constipation - Castor oil (1); 12-16 Senokots per week (1); eat bananas and avocados pears (1); laxatives (2)

Diarrhoea - medication (2); self-raising flour

Smearing - toilet paper in underwear (2)

Excessive flatus - Enos (1)

Dribbling urine - urinate frequently and wipe tip of penis

(ii) Coping/treatment methods

Constipation - Castor oil (1); 12-16 Senokots per week (1); eat bananas and avocado pears (1); laxatives (2)

Diarrhoea - medication (2); self-raising flour

Smearing - toilet paper in underwear (2)

Excessive flatus - Enos (1)

Dribbling urine - urinate frequently and wipe tip of penis